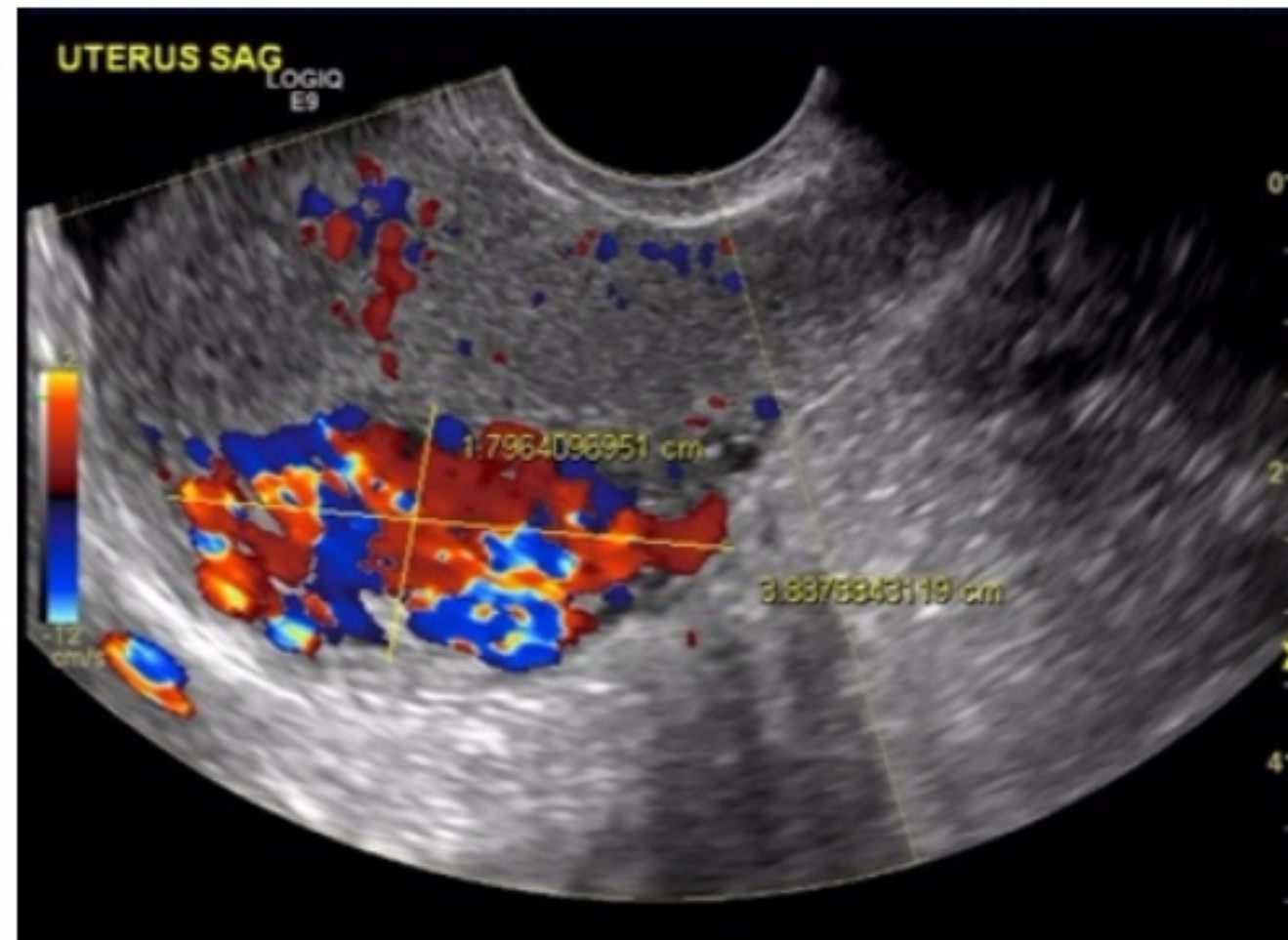
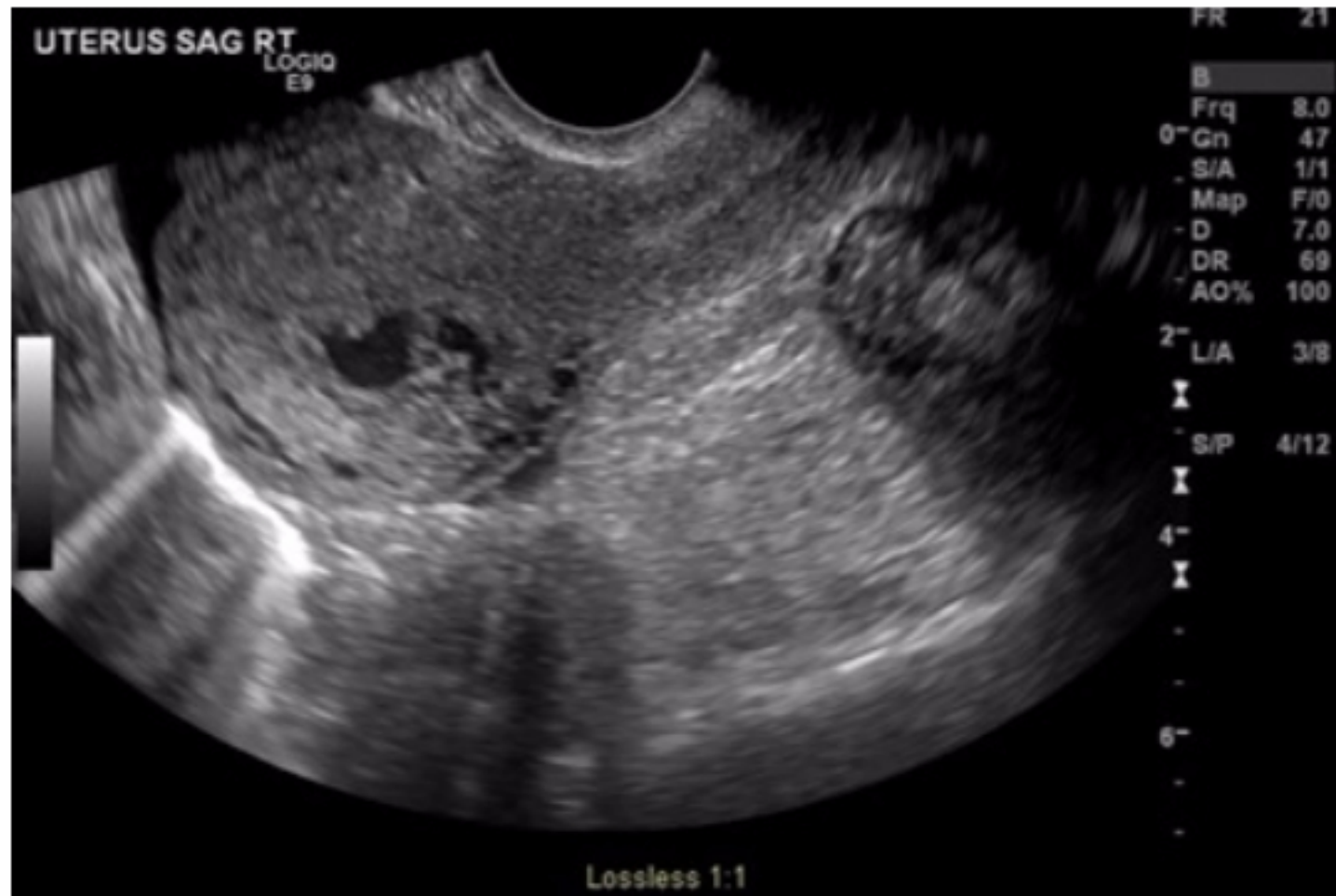


**History:** A 30-year-old woman presents to the emergency department with abnormal uterine bleeding. Her last menstrual period was nine days ago. Her past medical history includes an aborted pregnancy three months ago via dilation and curettage.

A pelvic ultrasound scan with Doppler was performed. Click images below to enlarge.



UTERUS SAG RT  
LOGIQ  
E9



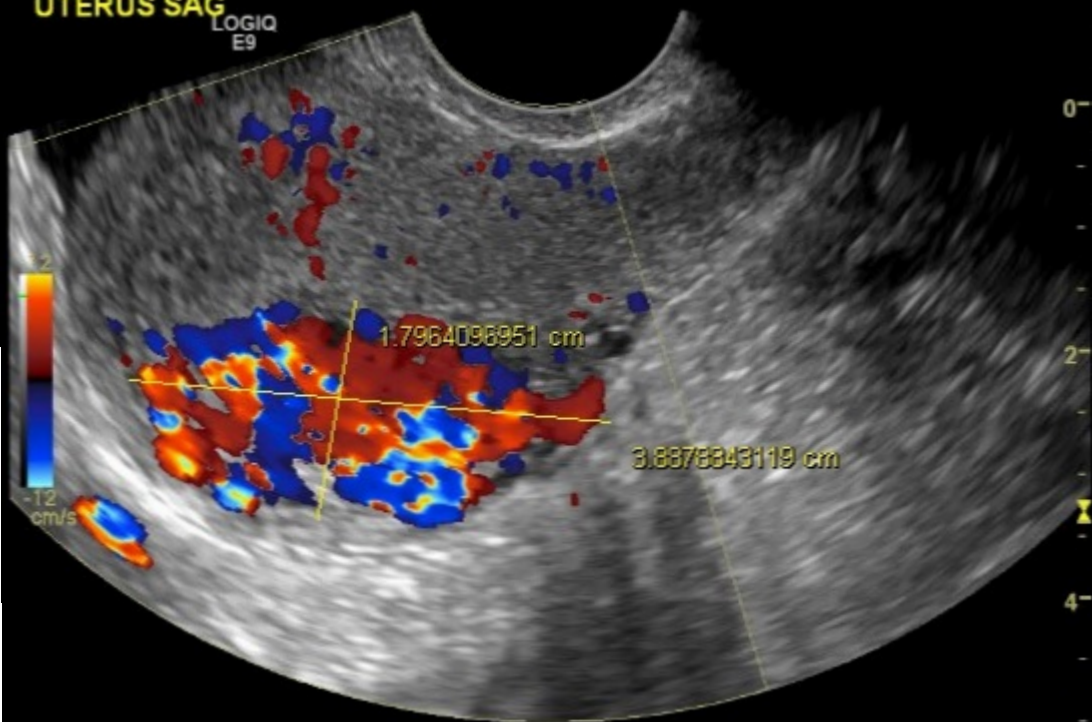
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- S/A	1/1
- Map	F/0
- D	7.0
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- AO%	100
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- S/P	4/12
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Lossless 1:1



UTERUS SAG

LOGIQ  
E9



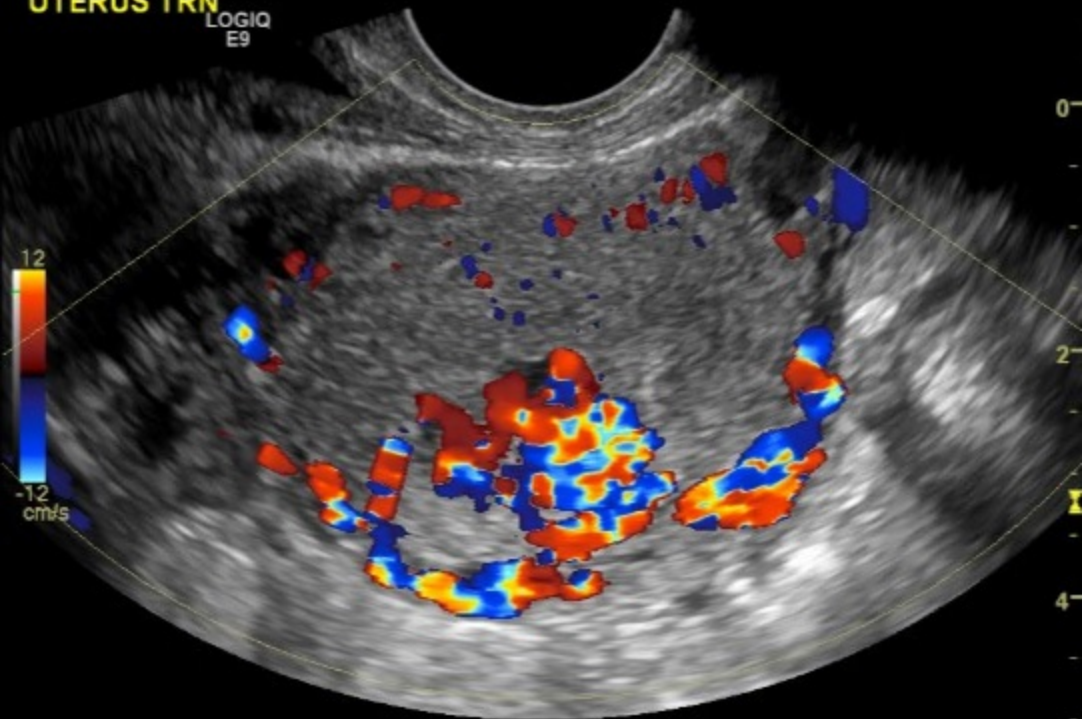
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3.8876643119 cm

-12  
cm/s

UTERUS TRN  
LOGIQ  
E9

12  
-12  
cm/s



**What is the finding from the pelvic ultrasound?**

- ☐ Intramural high-flow structure
- ☐ Endometrial high-flow structure
- ☐ Retained products of conception
- ☐ Leiomyoma

The question above accounts for 25% of your total score for this case.



**What is the finding from the pelvic ultrasound?**

☒ Intramural high-flow structure (correct!)

☐ Endometrial high-flow structure

☐ Retained products of conception

☐ Leiomyoma

The question above accounts for 25% of your total score for this case.

The question above accounts for 25% of your total score for this case.

**What is the next best imaging study to obtain for this ultrasound finding?**

☐ Pelvic CT angiography

☒ Pelvic MR angiography

☐ Hysterosalpingogram

The question above accounts for 20% of your total score for this case.

The question above accounts for 25% of your total score for this case.

**What is the next best imaging study to obtain for this ultrasound finding?**

☐ Pelvic CT angiography

☒ Pelvic MR angiography (correct!)

☐ Hysterosalpingogram

**[Explain this Answer]**

The question above accounts for 20% of your total score for this case.



☐ Retained products of conception

☐ Leiomyoma



MRI is the best confirmation imaging test due to its ability to differentiate soft tissues in the pelvis.

The question above

**What is the next**

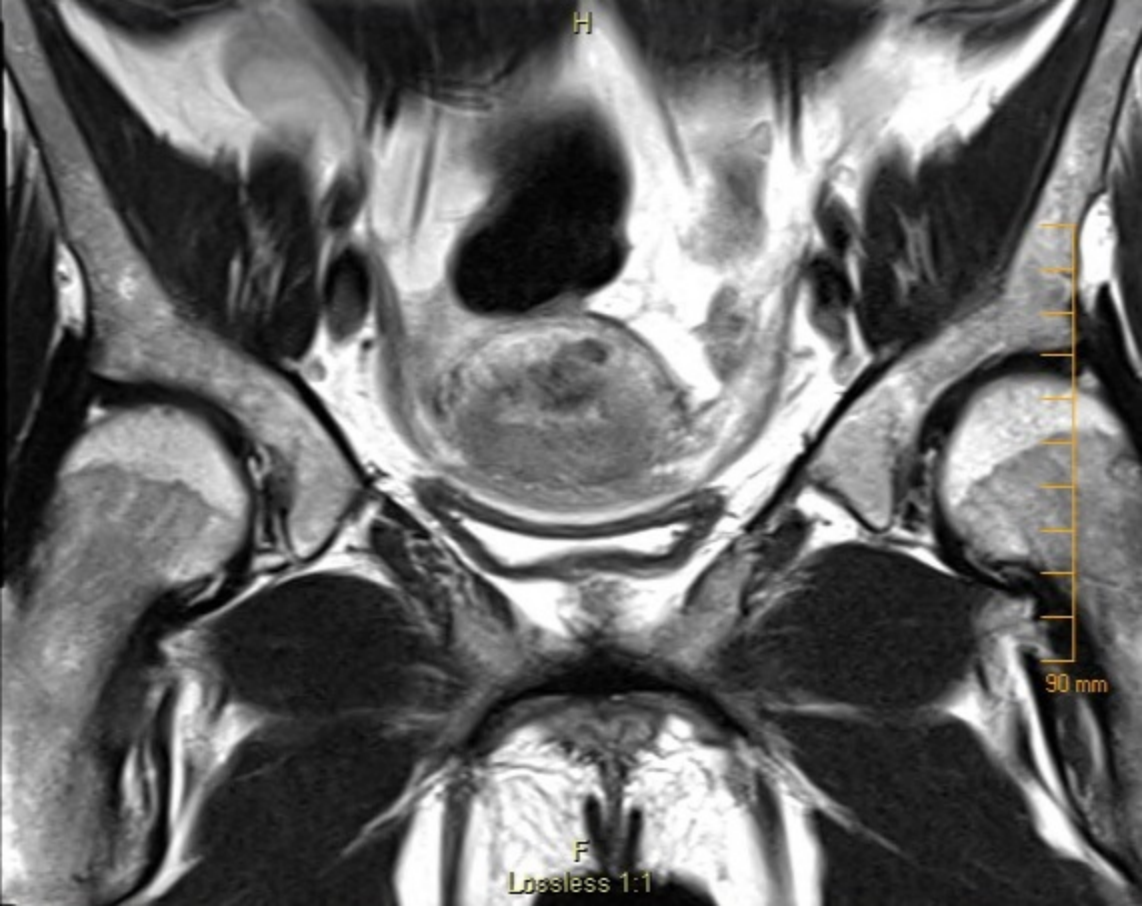
**and finding?**

☐ Pelvic CT ang

☒ Pelvic MR an



F  
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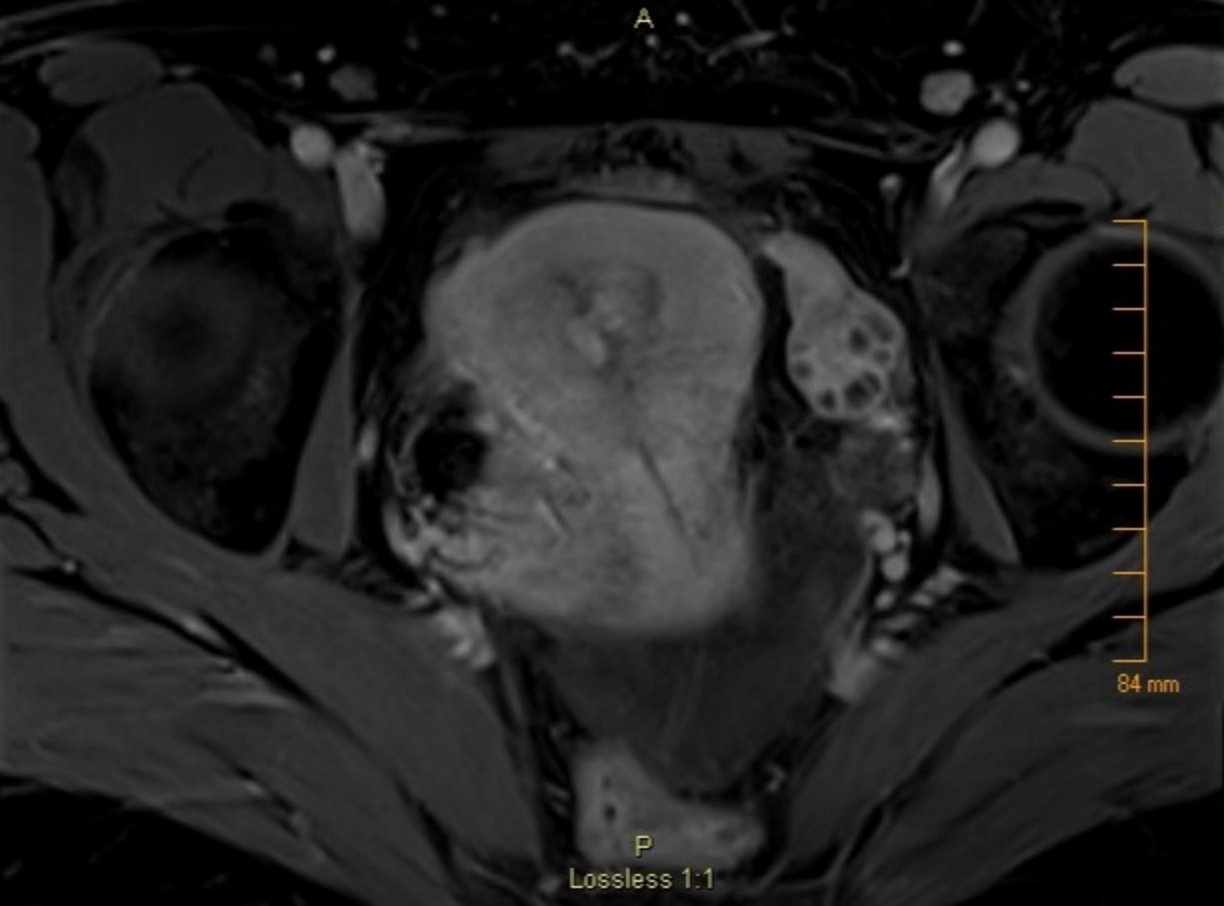
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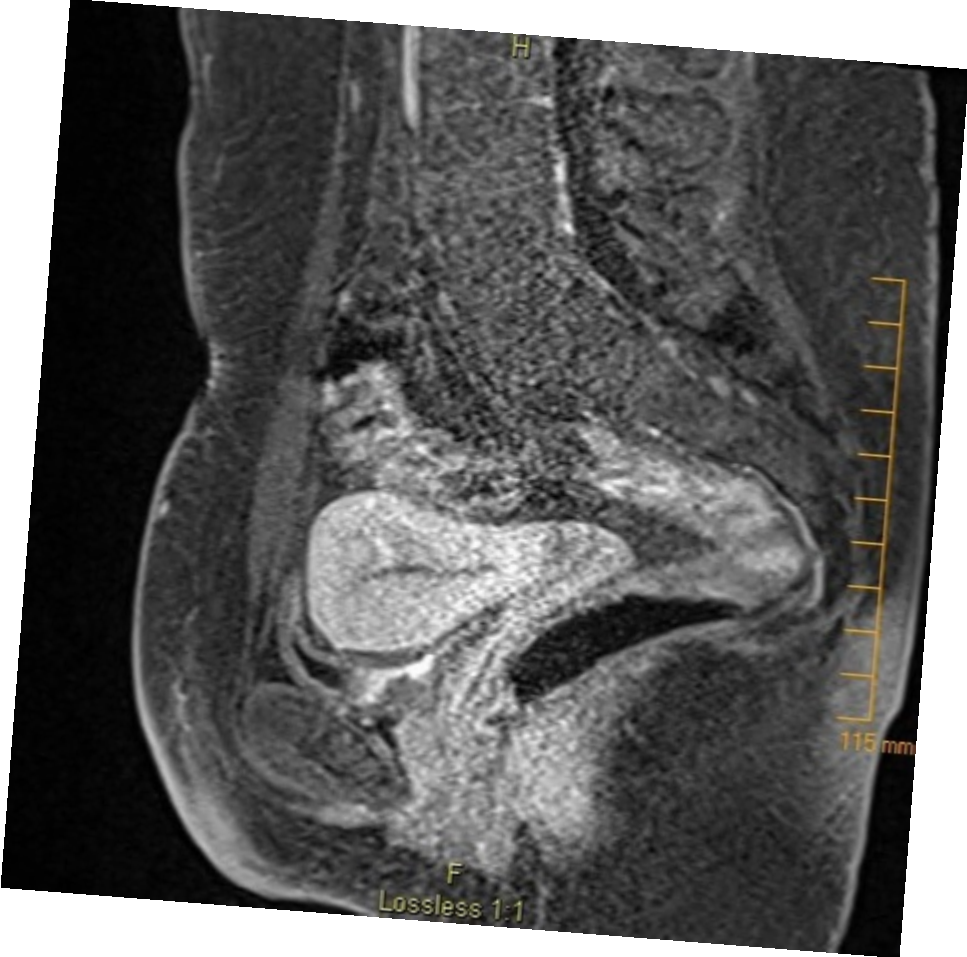
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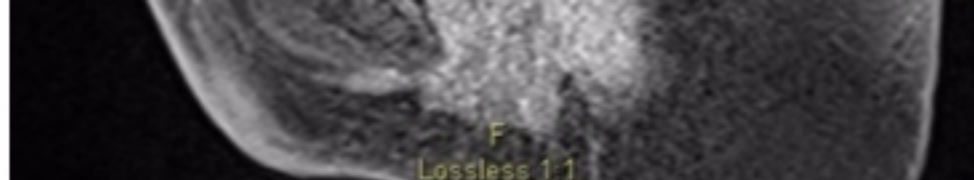
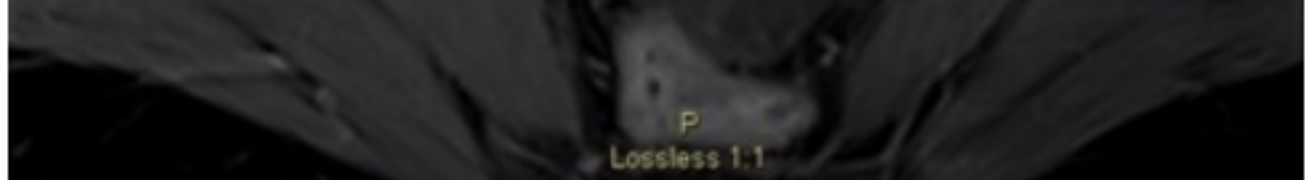
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Lossless 1:1









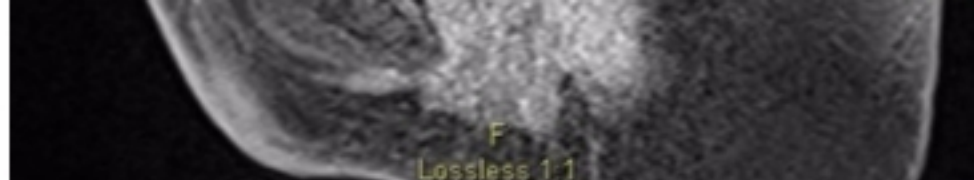
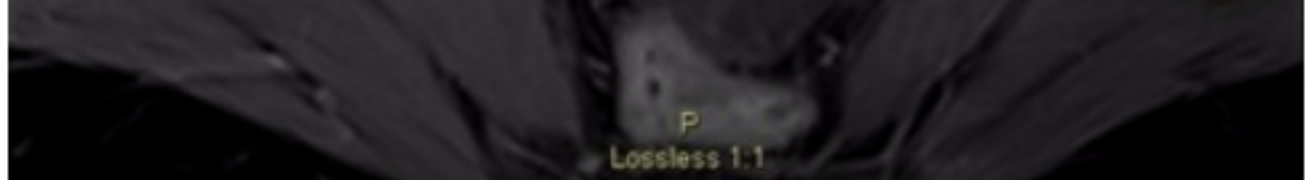
**Based on the MR findings, what is the next best step for the management of this patient?**

☐ Conservative management

☐ Myomectomy

☐ Uterine artery embolization

The question above accounts for 25% of your total score for this case.



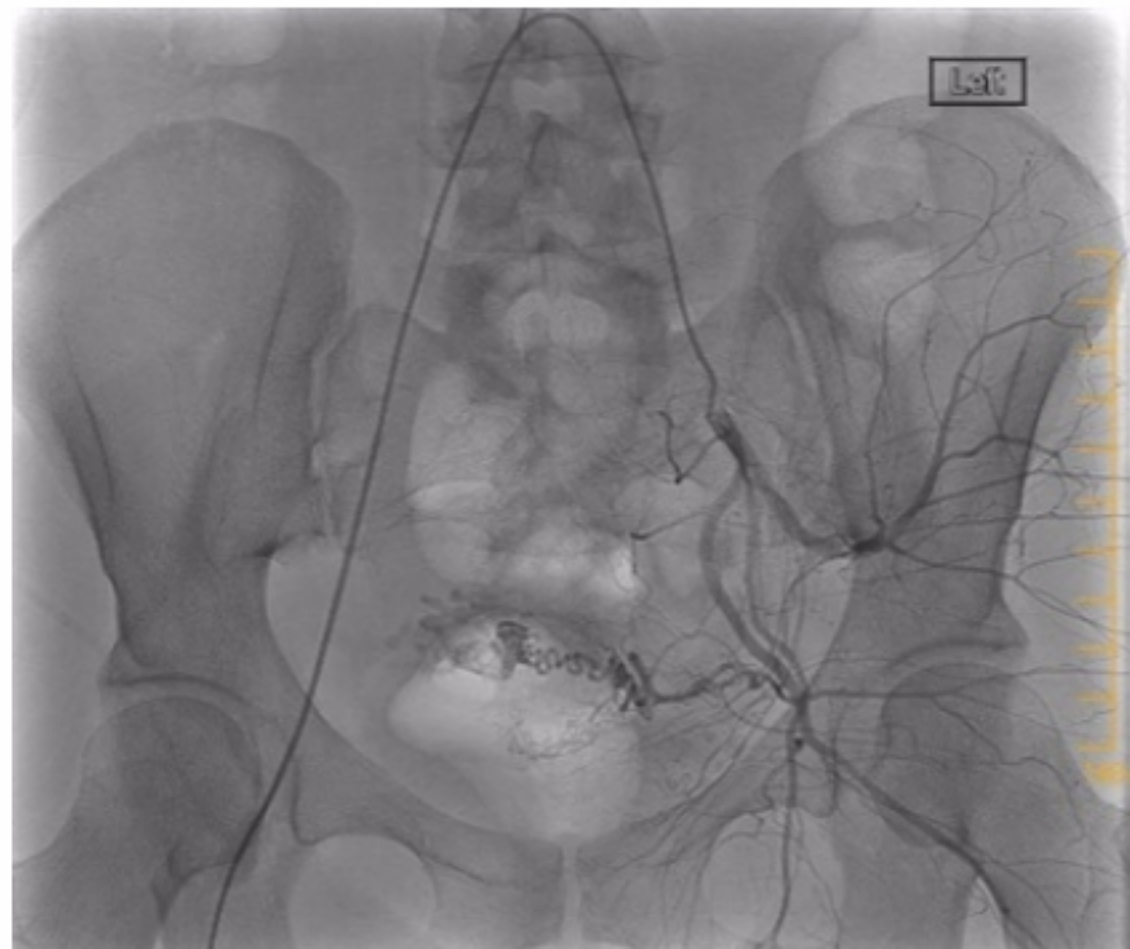
**Based on the MR findings, what is the next best step for the management of this patient?**

- ☐ Conservative management
- ☐ Myomectomy
- ☒ Uterine artery embolization (correct!)

The question above accounts for 25% of your total score for this case.

## Angiography images

The patient was brought to the interventional radiology suite for pelvic angiography. Gelfoam was used to embolize the uterine arteries and stop uterine bleeding.





Left



Let





## Findings

- **Ultrasound:** There is a 3.9 x 2.7 x 1.8-cm markedly vascular tissue possibly representing an arteriovenous malformation (AVM) within the uterus involving the posterior fundus central and right with some left-sided involvement. The vascular tissue extends from external wall all the way to the submucosal endometrial layer.
- **MRI:** Focal serpiginous flow voids are centered within the posterosuperior myometrium of the uterus with extension into the endometrial canal with associated mild enhancement.
- **Angiography:** There is an abnormal area of vascularity in the uterine region with early draining venous structures.

**Diagnosis:** Uterine arteriovenous malformation secondary to uterine instrumentation

## Additional questions

**Is this diagnosis more common in premenopausal or postmenopausal women?**

☐ Premenopausal

☐ Postmenopausal

The question above accounts for 15% of your total score for this case.

**Which of the following is NOT associated with the development of uterine AVMs?**



## Additional questions

Is this diagnosis more common in premenopausal or postmenopausal women?

☒ Premenopausal (correct!)

☐ Postmenopausal

[**Explain this Answer**]

The question above accounts for 15% of your total score for this case.

Which of the following is NOT associated with the development of uterine

**Which of the following is NOT associated with the development of uterine AVMs?**

- ☐ Gestational trophoblastic disease
- ☐ Dilation and curettage
- ☐ Pelvic inflammatory disease
- ☐ Hereditary hemorrhagic telangiectasia

The question above accounts for 15 % of your total score for this case.

**Which of the following is NOT associated with the development of uterine AVMs?**

- ☐ Gestational trophoblastic disease
- ☐ Dilation and curettage
- ☒ Pelvic inflammatory disease (correct!)
- ☐ Hereditary hemorrhagic telangiectasia

## Intrauterine arteriovenous malformations (AVMs)

- Intrauterine AVMs are an uncommon source of abnormal uterine bleeding.
- About half of cases are congenital and can present with no prior history of surgery or trauma.
- The other half of cases occur in premenopausal women due to hormonal changes in pregnancy, trauma during labor pregnancy, or surgical trauma.
- If the patient's condition allows, fast-flow uterine malformations that are discovered on pelvic ultrasound should be followed up with MR angiography to determine the full extent and nature of the lesion.
- Percutaneous embolization is a safe and effective treatment for arteriovenous malformations and preserves uterine function in patients who wish to have children in the future. In this case, Gelfoam was used to embolize the uterine arteries and stop



uterine bleeding.

- Although Gelfoam was used in this case, there is no consensus for the best embolic agent to use for treating uterine arteriovenous malformations.

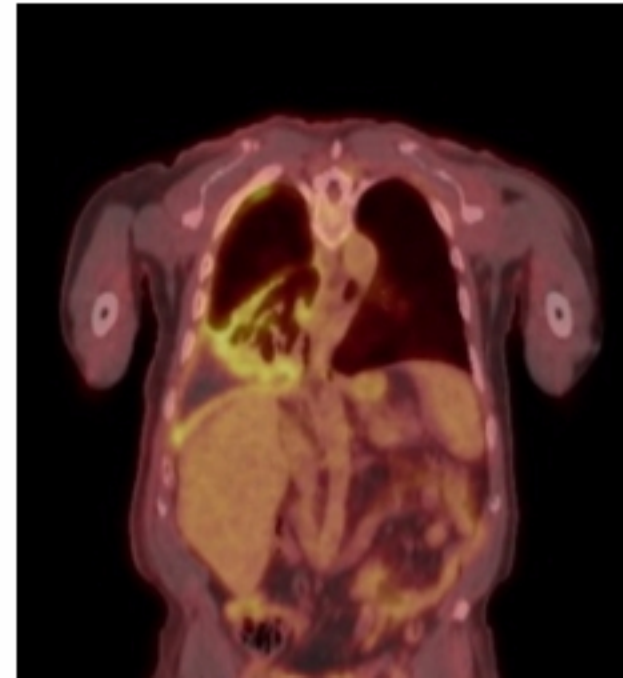
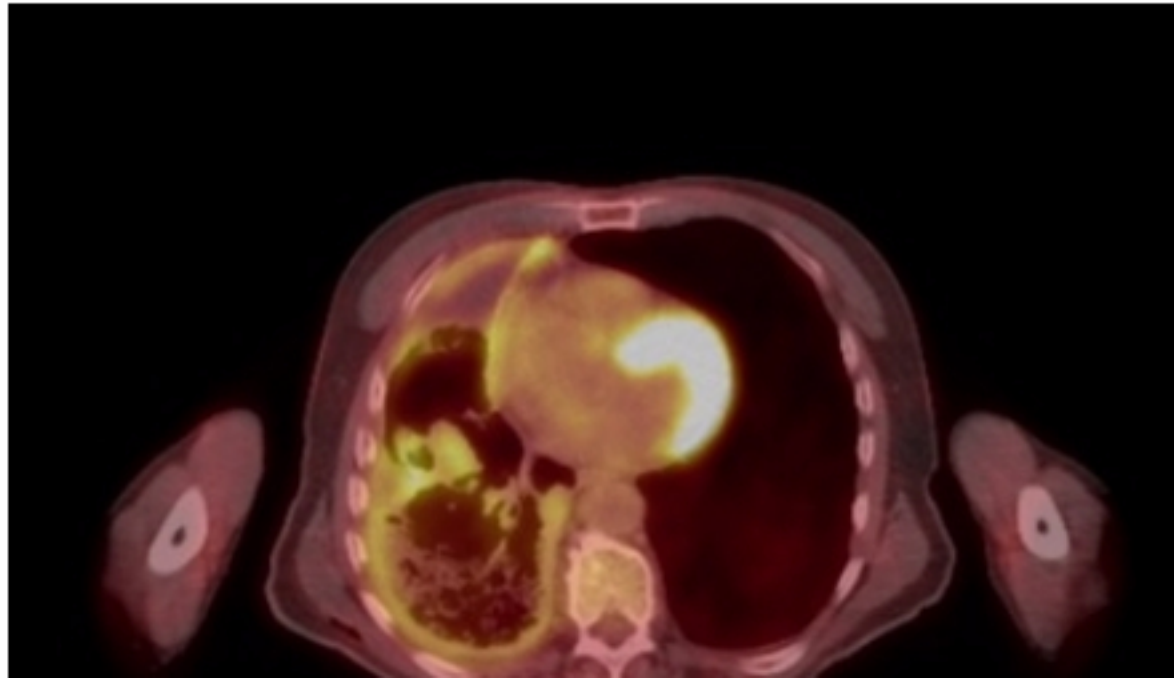
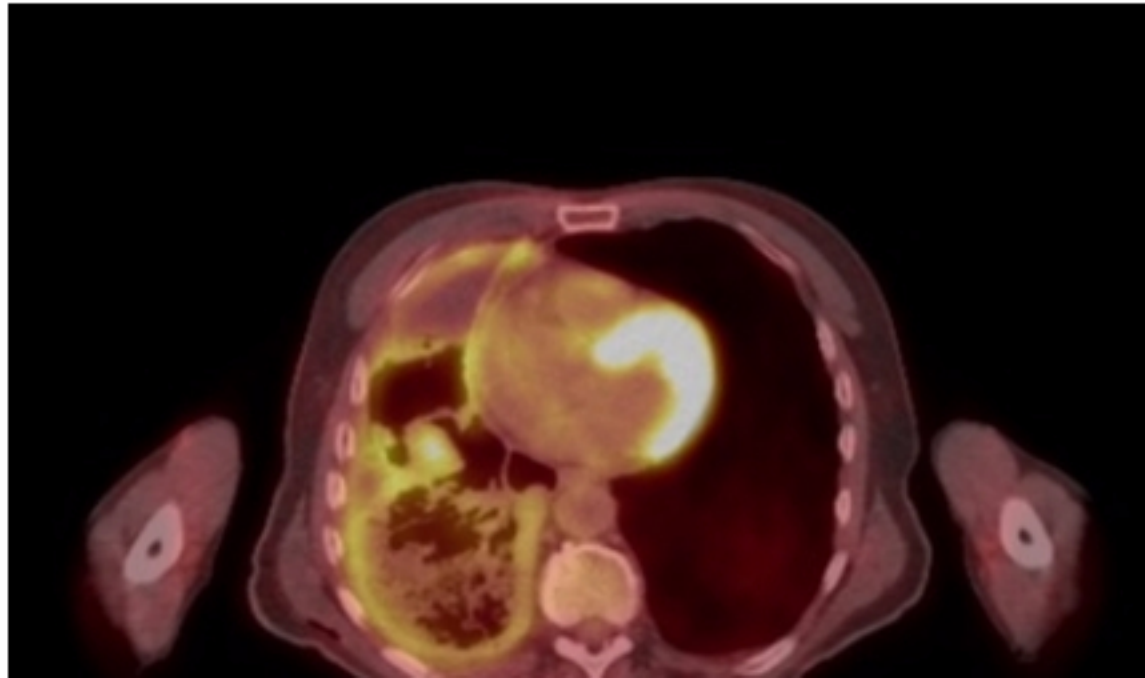
## References

1. Burrows PE. Vascular malformations involving the female pelvis. *Semin Intervent Radiol*. 2008;25(4):347-360.
2. Ghai S, Rajan DK, Asch MR, Muradali D, Simons ME, TerBrugge KG. Efficacy of embolization in traumatic uterine vascular malformations. *J Vasc Interv Radiol*. 2003;14(11):1401-1408.
3. Peitsidis P, Manolakos E, Tsekoura V, Kreienberg R, Schwentner L. Uterine arteriovenous malformations induced after diagnostic curettage: a systematic review. *Arch Gynecol Obstet*. 2011;284(5):1137-1151.
4. Wang Z, Chen J, Shi H, et al. Efficacy and safety of embolization in iatrogenic traumatic



**History:** A 79-year-old man initially presented to his primary care provider two years ago complaining of a cough and shortness of breath. At that time, the patient underwent chest radiography, which was suspicious for pleural thickening. A PET/CT scan was recommended for further evaluation.

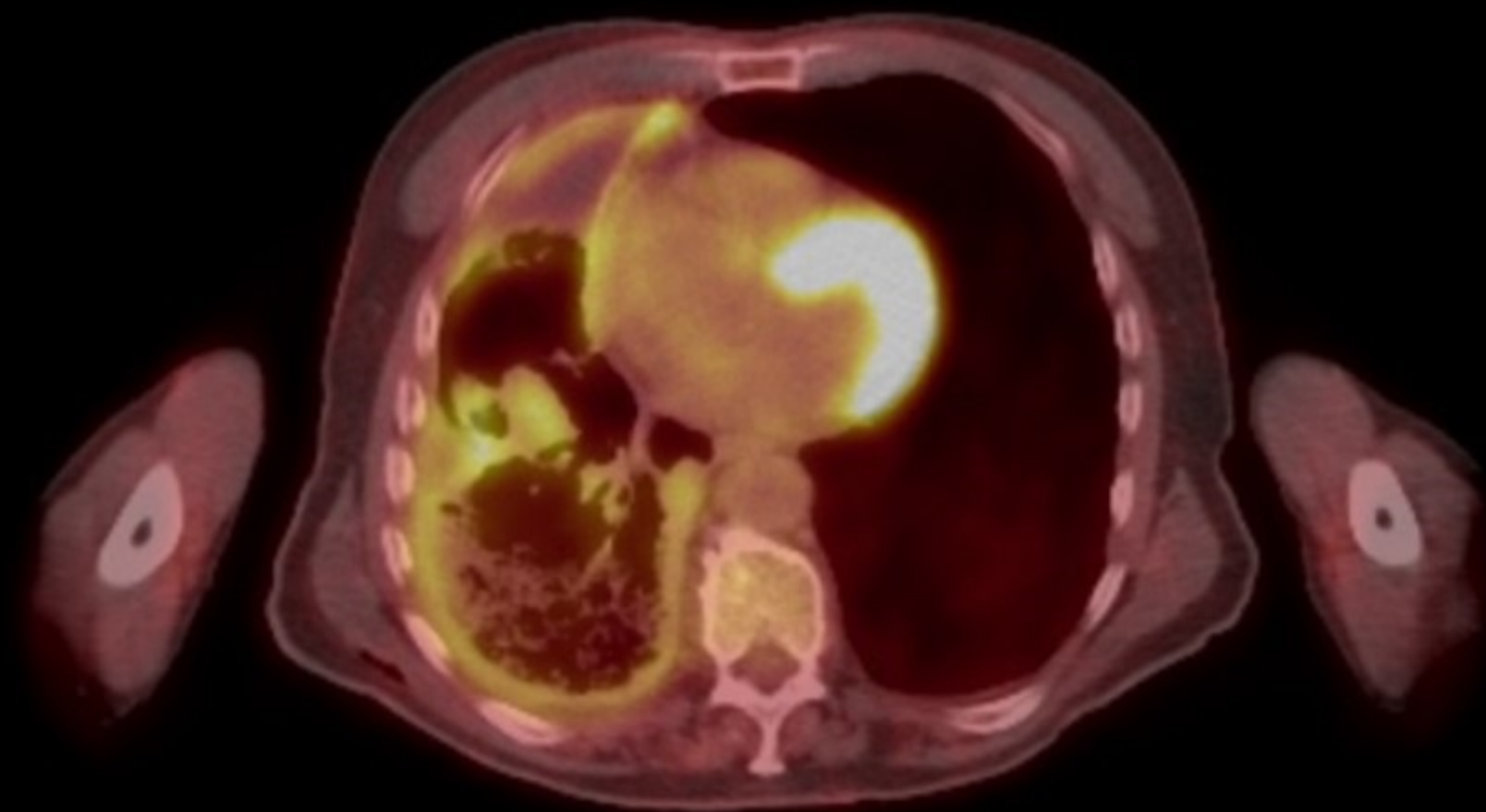
Axial, coronal, and sagittal views of the fused PET/CT images of the chest are shown below. Click to enlarge.





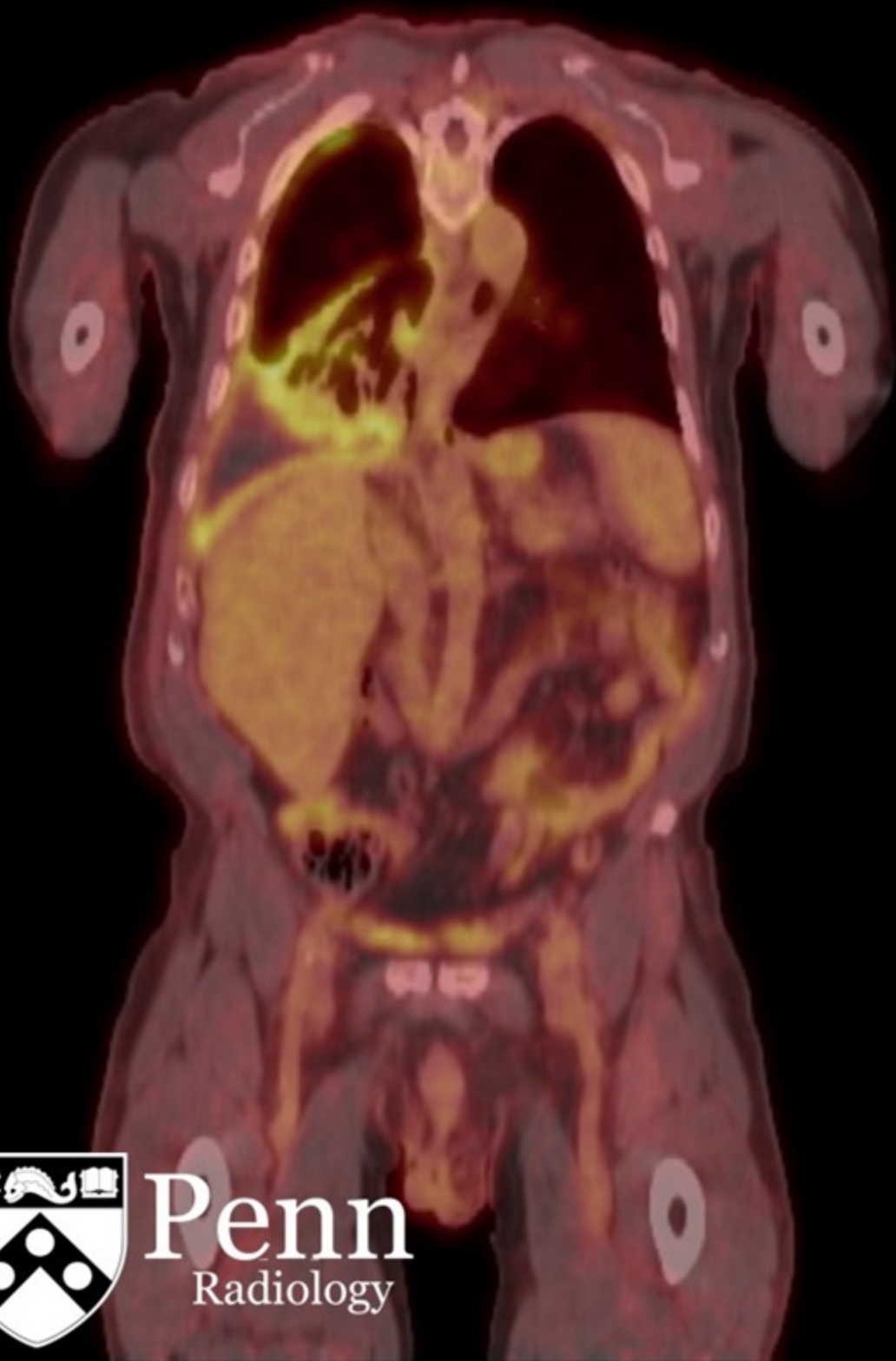
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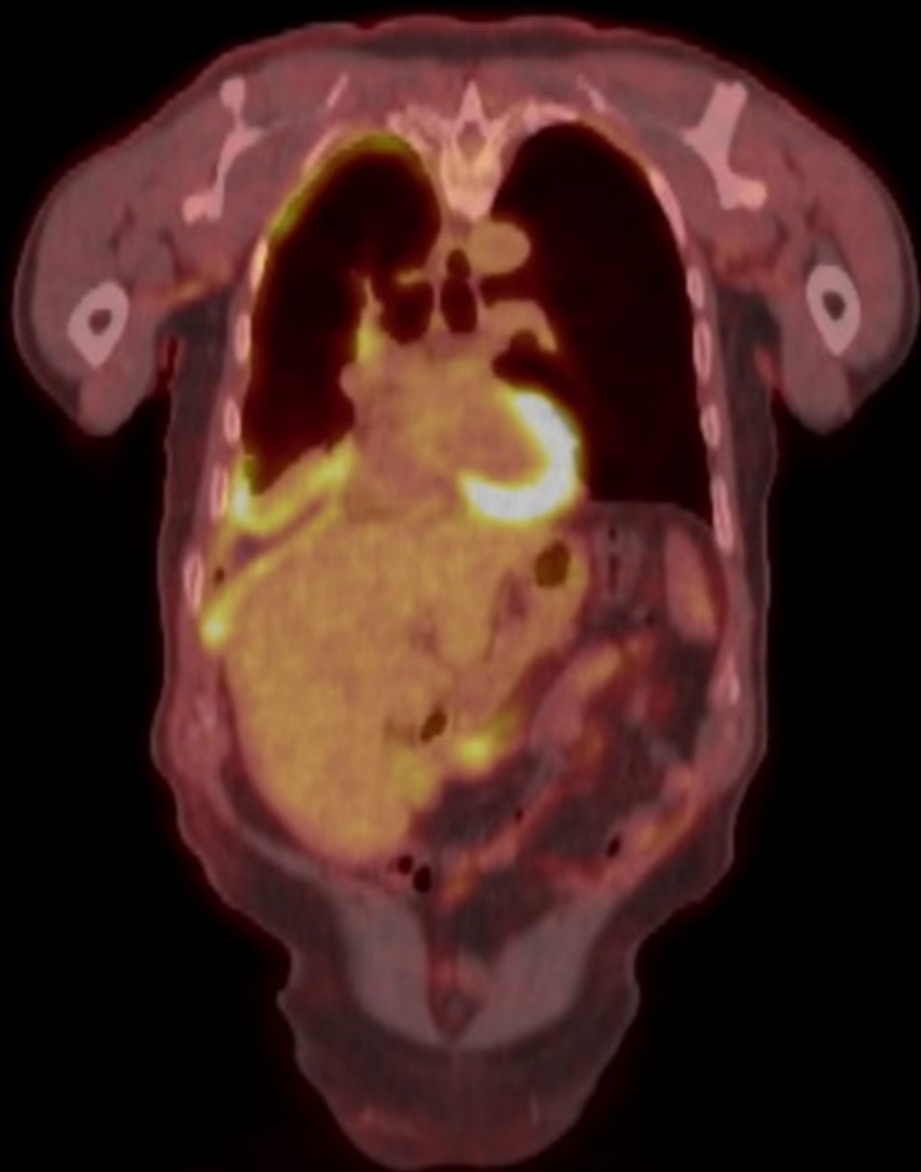
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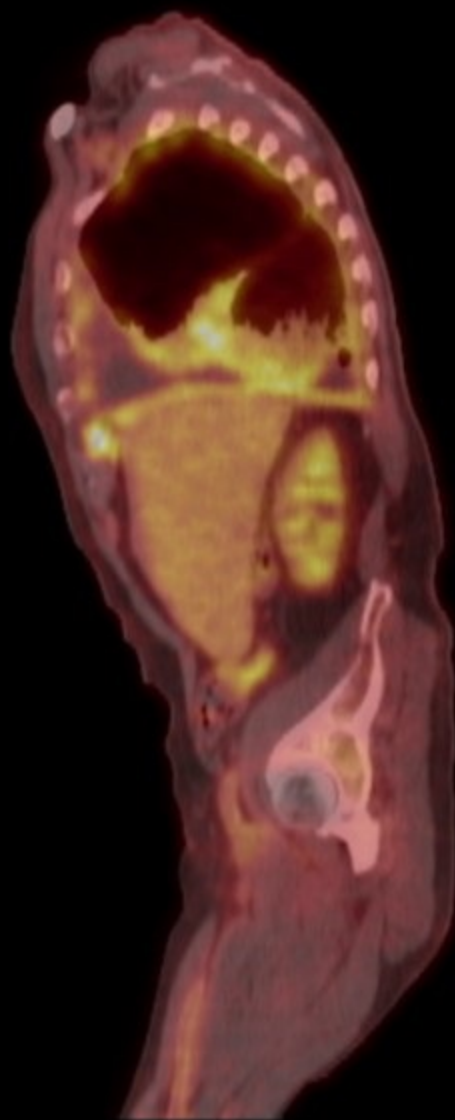
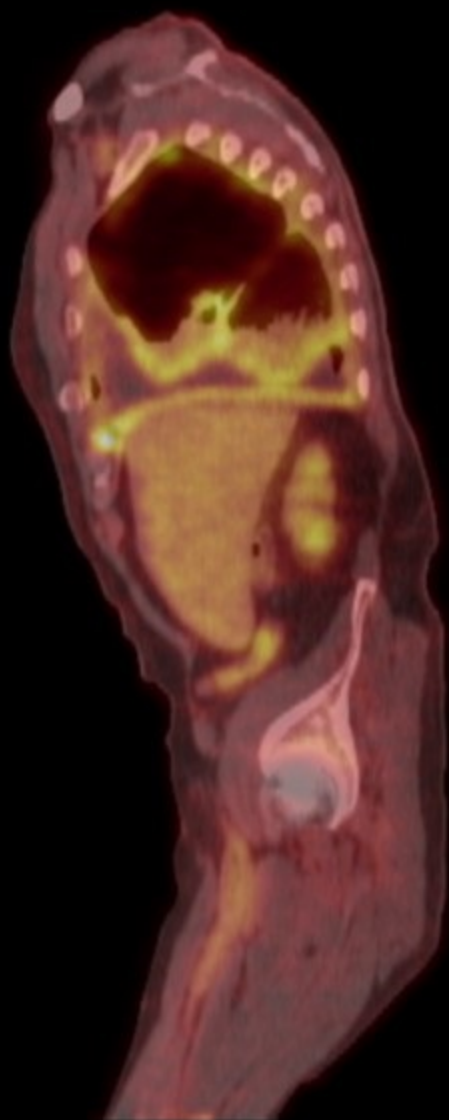


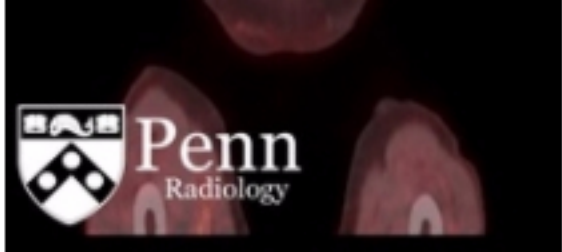
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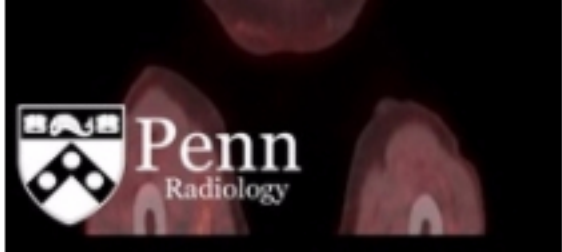
**There is normal, physiologic uptake of F-18 FDG.**

☐ True

☐ False

The question above accounts for 16% of your total score for this case.

[VIEW YOUR SCORE](#)



**There is normal, physiologic uptake of F-18 FDG.**

☐ True

☒ False (correct!)

The question above accounts for 16% of your total score for this case.

[VIEW YOUR SCORE](#)

## Chest CT images

PET/CT showed diffuse right pleural thickening, which appears more nodular in appearance along the major fissure. This may be inflammatory or neoplastic. Close interval follow-up was recommended.

The patient was instructed to follow-up in three months but was lost to follow-up. Two years later, the patient presented with worsening shortness of breath, continued weight loss, and back pain. An unenhanced CT scan of the chest was ordered.

Axial, coronal, and sagittal views from the chest CT scan are shown below. Click to enlarge.

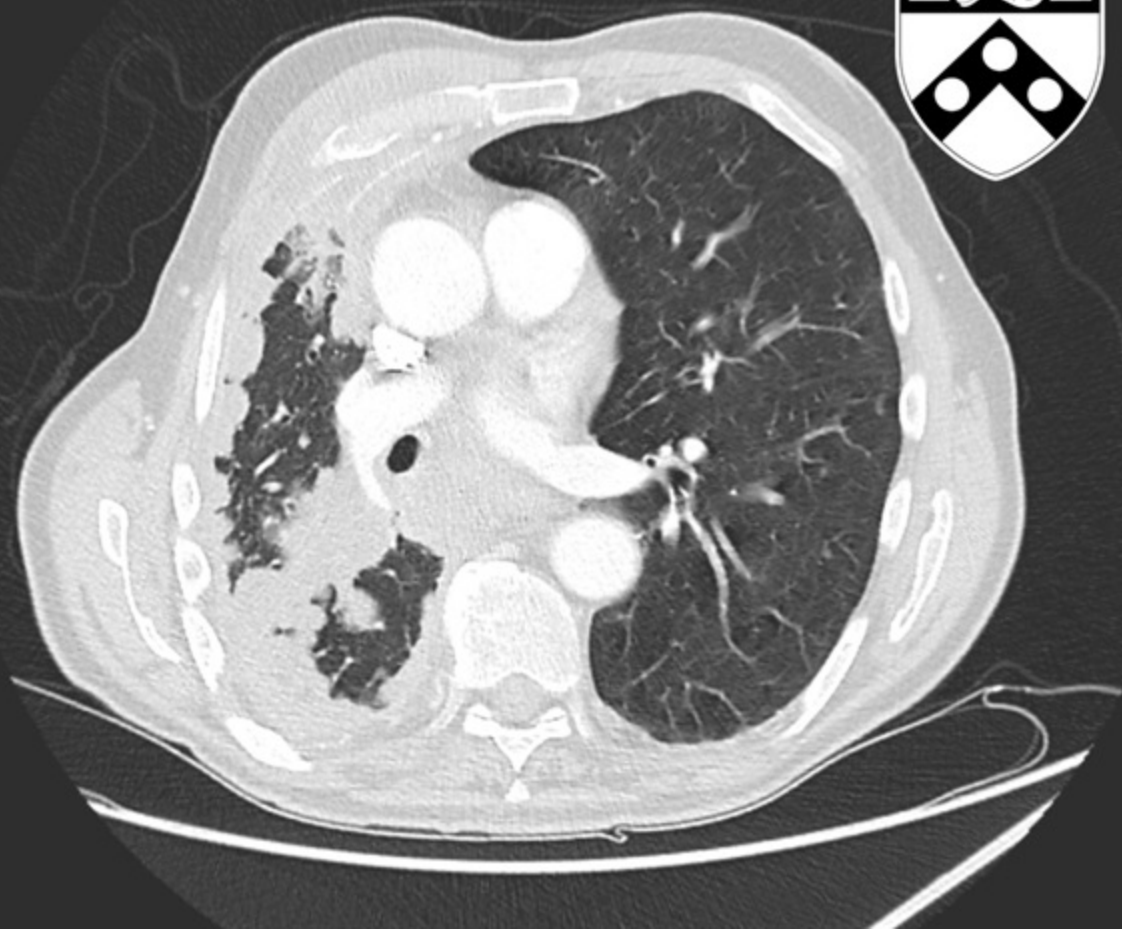






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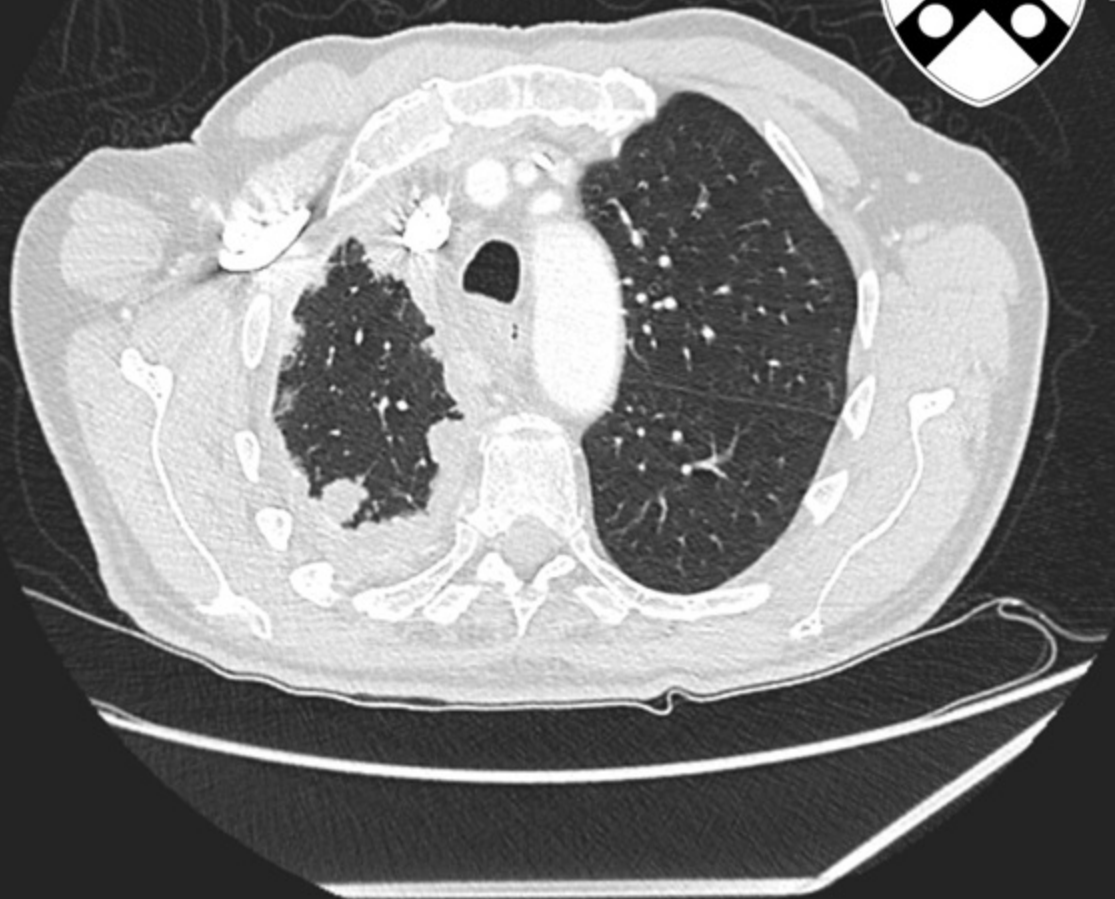
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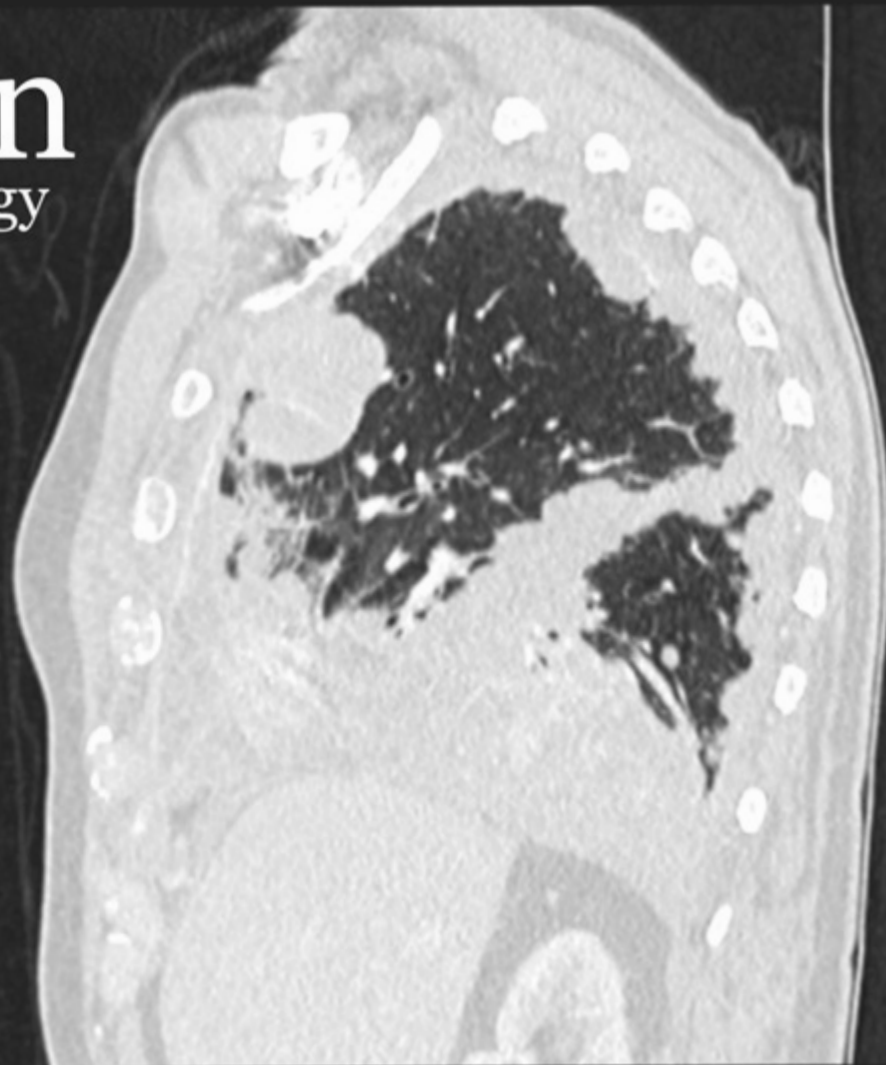


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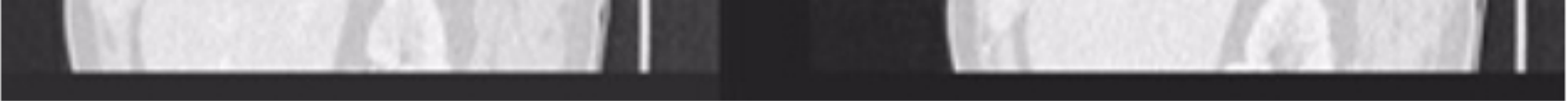
**Based on the chest CT and PET/CT scans from two years prior, what is the best diagnosis to explain the image findings and the patient's complaints?**

☐ Chronic pleural effusion

☐ Subpleural fat

☐ Diffuse large B-cell lymphoma

☐ Malignant mesothelioma



**Based on the chest CT and PET/CT scans from two years prior, what is the best diagnosis to explain the image findings and the patient's complaints?**

☐ Chronic pleural effusion

☐ Subpleural fat

☐ Diffuse large B-cell lymphoma

☒ Malignant mesothelioma (correct!)

## Findings

- **PET/CT:**
  - There is intense diffuse right pleural uptake associated with intense bilateral ill-defined hilar lymph node uptake.
  - There is diffuse pleural thickening, more nodular in appearance along the major fissure, nonspecific, may be inflammatory or neoplastic. Close interval follow-up was recommended at the time.
- **Chest CT:** There is circumferential and nodular pleural thickening in the right costal, mediastinal, and diaphragmatic pleural surfaces, in keeping with pleural mesothelioma. Pleural thickening is most pronounced along the diaphragmatic pleura and within the fissures.

## Differential diagnosis

## Differential diagnosis

- Malignant pleural mesothelioma
- Chronic pleural effusion/empyema
- Primary synovial sarcoma of lung
- Reactive mesothelial cell hyperplasia
- Organizing pleuritis

**Diagnosis:** Malignant right-sided pleural mesothelioma with transdiaphragmatic disease and evidence of chest wall invasion

[VIEW YOUR SCORE](#)



## 79-year-old man with cough, shortness of breath

CASE OUTLINE

Page 4 of 5

### Additional questions

**Mesothelioma is the most common primary malignancy of the pleura.**

☐ True

☐ False

The question above accounts for 16% of your total score for this case

## 79-year-old man with cough, shortness of breath

CASE OUTLINE

Page 4 of 5

### Additional questions

**Mesothelioma is the most common primary malignancy of the pleura.**

☒ True (correct!)

☐ False

The question above accounts for 16% of your total score for this case

## What is the primary treatment modality of malignant pleural mesothelioma?

- ☐ A. Extrapleural pneumonectomy
- ☐ B. Pleurectomy and decortication
- ☐ C. Chemotherapy
- ☐ D. External-beam radiation therapy
- ☐ E. None of the above
- ☒ F. Both A and B (correct!)



Surgery is the mainstay of therapy, and local control is important for long-term survival.

[Explain this Answer]

The question above

Which type of

☐ Epithelioid

st prognosis?



[[Explain this Answer](#)]

The question above accounts for 16% of your total score for this case.

**Which type of mesothelioma subtype is associated with the best prognosis?**

☒ Epithelioid

☐ Sarcomatoid

☐ Mixed

The question above accounts for 16% of your total score for this case.

**Exposure to which of the following substances is most closely associated**

[[Explain this Answer](#)]

The question above accounts for 16% of your total score for this case.

**Which type of mesothelioma subtype is associated with the best prognosis?**

☒ Epithelioid (correct!)

☐ Sarcomatoid

☐ Mixed

The question above accounts for 16% of your total score for this case.

**Exposure to which of the following substances is most closely associated**

**Exposure to which of the following substances is most closely associated with the development of mesothelioma?**

☒ Polychlorinated biphenyls (PCBs)

☐ Dioxins

☐ Asbestos

☐ Phthalates

The question above accounts for 16% of your total score for this case

**Exposure to which of the following substances is most closely associated with the development of mesothelioma?**

☐ Polychlorinated biphenyls (PCBs)

☐ Dioxins

☒ Asbestos (correct!)

☐ Phthalates

The question above accounts for 16% of your total score for this case



# Malignant pleural mesothelioma

## Pathophysiology

Malignant pleural mesothelioma is the most common primary neoplasm of the pleura.

- It has a strong association with asbestos exposure, particularly crocidolite.
- It can develop in patients with transient or indirect exposure to asbestos; a dose-dependent relationship has been recognized.
- It can occur in the pleura, peritoneum, pericardium, or tunica vaginalis. Pleural mesothelioma represents 90% of disease.
- Simian virus 40 (SV40) has been associated with malignant mesothelioma and suggested as a causal cofactor.

## Epidemiology

- Simian virus 40 (SV40) has been associated with malignant mesothelioma and suggested as a causal cofactor.

## **Epidemiology**

- Approximately 80% of mesothelioma patients have prior asbestos exposure.
- The latency period can be more than 40 years.
- An estimated 2,500 to 3,500 cases occur per year in the U.S., predominantly in elderly men.
- It is most commonly seen in patients 60 years and older.
- Highest annual rates of disease reported in Australia and the U.K.
- The risk of disease increases with age and is higher in men.
- With the exception of the U.S., worldwide incidence continues to increase.

## **Clinical presentation**

The clinical presentation is nonspecific and is usually with the following:

## Clinical presentation

Typically, patients are 60 years and older with the following:

- Previous exposure to asbestos
- Insidious onset of nonspecific symptoms such as dyspnea, cough, and chest pain
- Systemic symptoms such as weight loss and fatigue
- Unilateral pleural effusions

Biopsy is required to confirm the diagnosis.

## Imaging features

- CT:
  - Pleural thickening is most common finding, can be lobular or smooth, with extension along the fissures
  - Pleural effusions are commonly seen



- Pleural thickening is most common finding, can be lobular or smooth, with extension along the fissures
- Pleural effusions are commonly seen.
- Contraction of the hemithorax, with or without mediastinal shift.
- Mesothelioma is not known to arise from pleural plaques.
- As disease advances, there is invasion of mediastinal structures including pericardium, great vessels and esophagus
- F-18 FDG PET:
  - F-18 uptake is significantly greater in malignant mesothelioma than benign pleural disease
  - Useful in detection of nodal disease
- Chest radiography:
  - Usually shows an effusion with possibly pleural thickening
  - Pleural thickening becomes more lobulated as tumor progresses
  - Contraction of the affected hemithorax

- Pleural thickening becomes more lobulated as tumor progresses
- Contraction of the affected hemithorax

## Treatment

- Surgery is the mainstay of therapy for malignant pleural mesothelioma.
- Pleurectomy and decortication are used for early disease.
- Extrapleural pneumonectomy (EPP) also is used.
- Adjuvant radiotherapy following pleurectomy is used for locoregional control.
- Chemotherapy with pemetrexed and cisplatin is used for patients who are fit enough to tolerate it.

## References

1. Cugell DW, Kamp DW. Asbestos and the pleura: A review. *Chest*. 2004;125(3):1103-1117.
2. Beach HD, Davies GJ, Attanoos R, Crane M, Adams H, Phillips S. Asbestos: When the

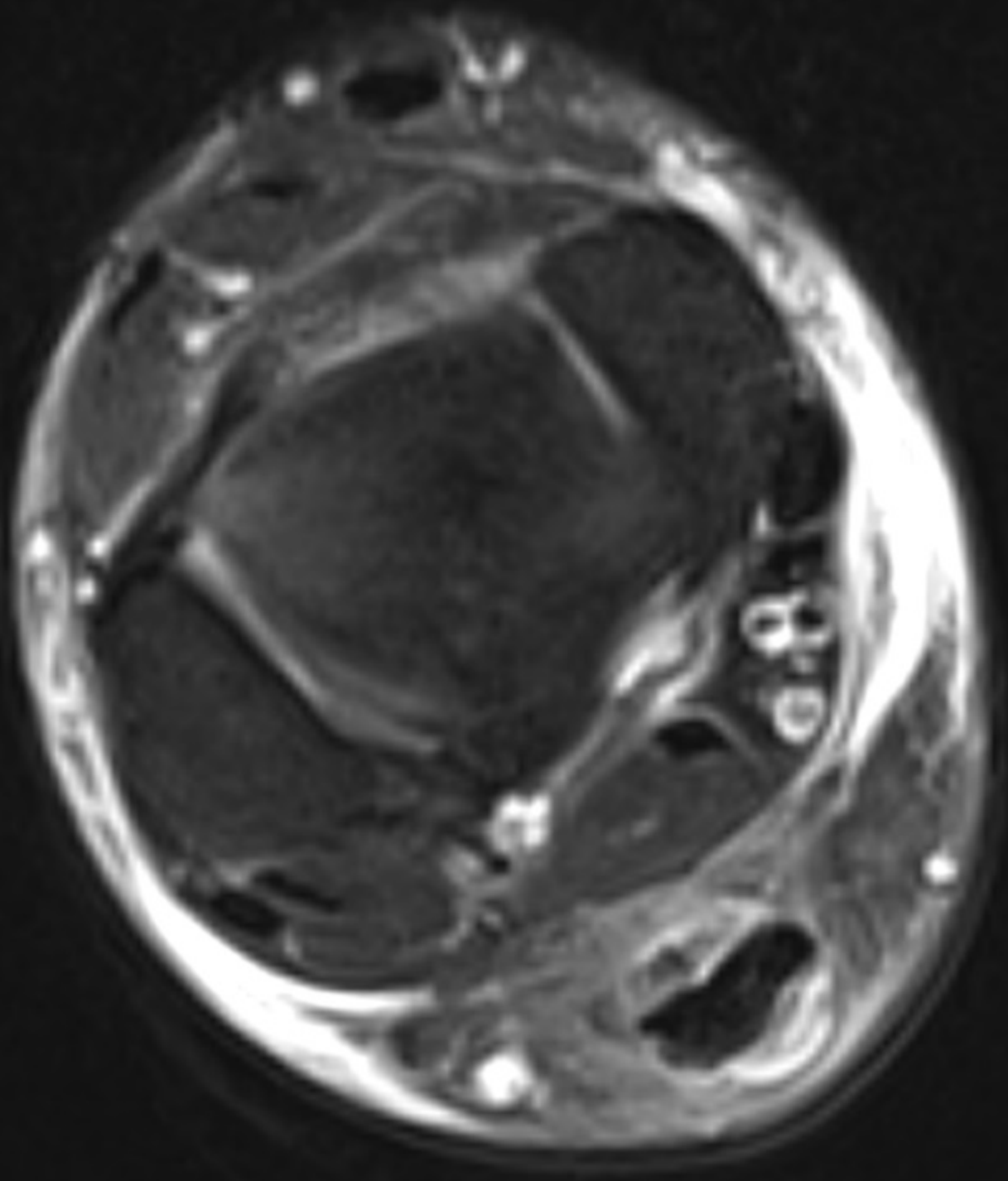


## History and MR images

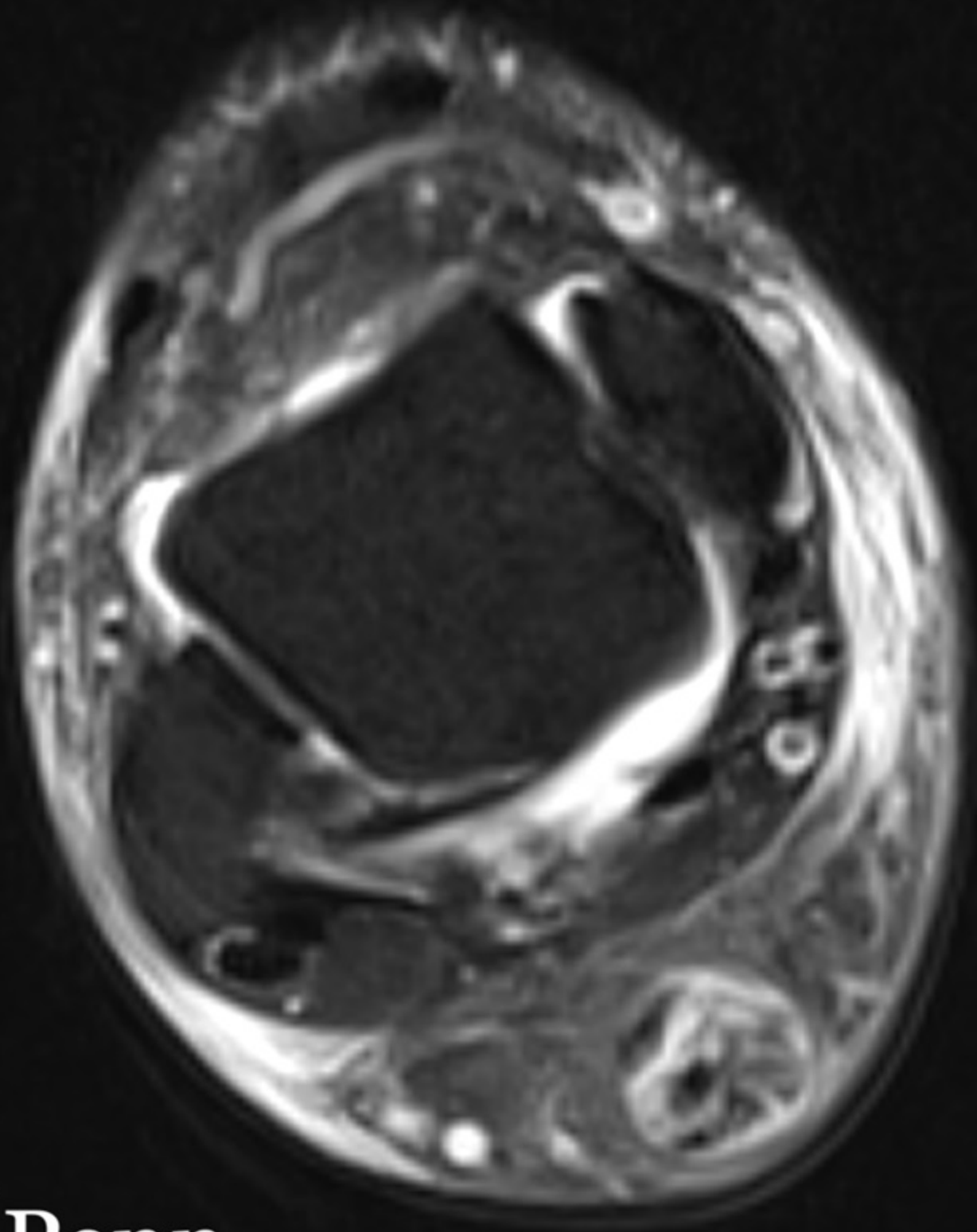
Our appreciation is extended to Dr. Teresa Martin-Carreras, University of Pennsylvania Department of Radiology, for contributing this case.

**History:** A 49-year-old man presents to the emergency department with a three-day history of right ankle pain, a new bump behind his ankle, and a "clicking/popping" sound following a run.

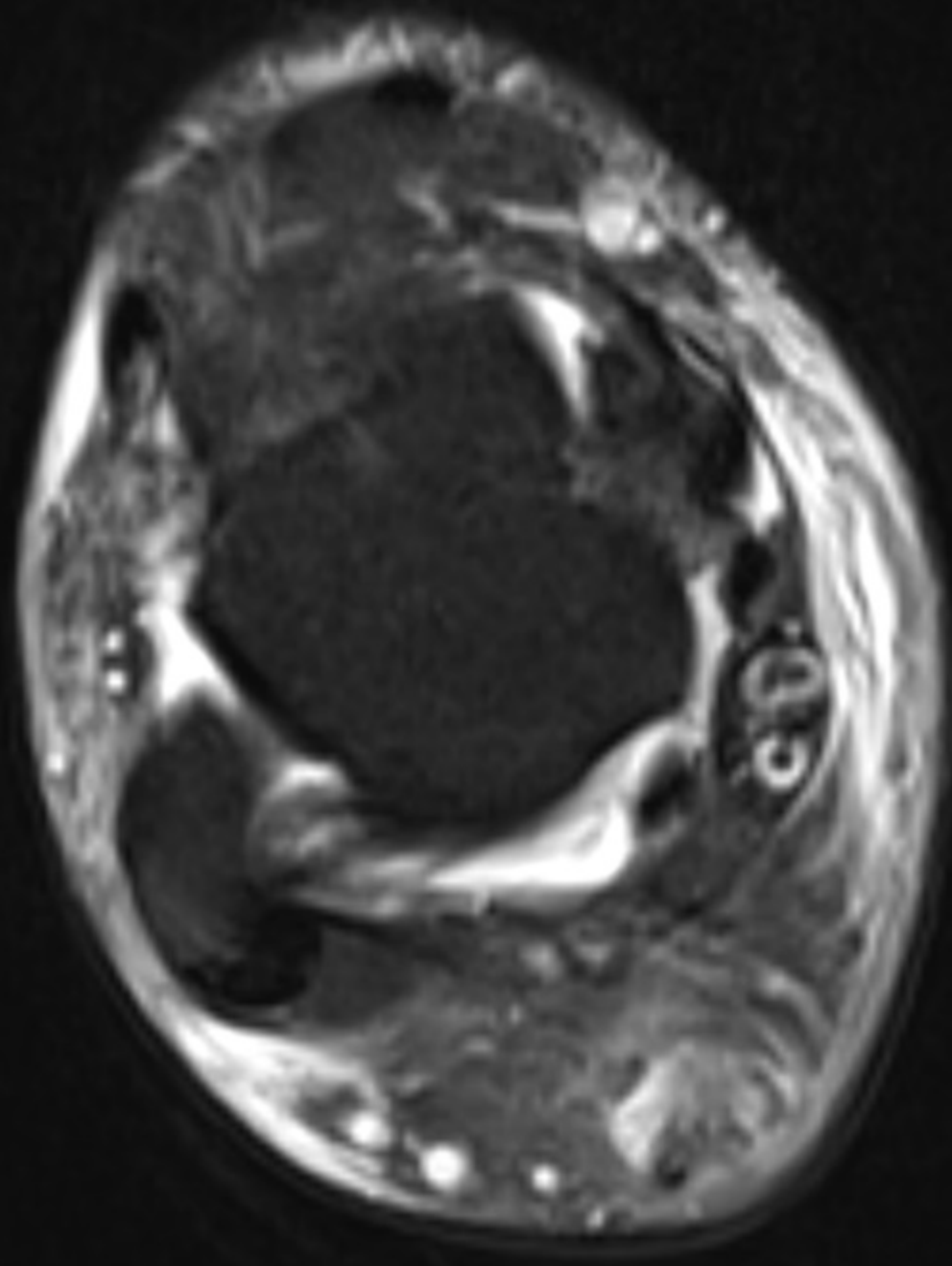
He subsequently underwent an MRI scan without contrast of the right calf. Select images are shown below. Click to enlarge. In order: axial T2-weighted fat-saturated (proximal to distal), sagittal short tau inversion-recovery (STIR), coronal STIR, and axial T1-weighted sequences.



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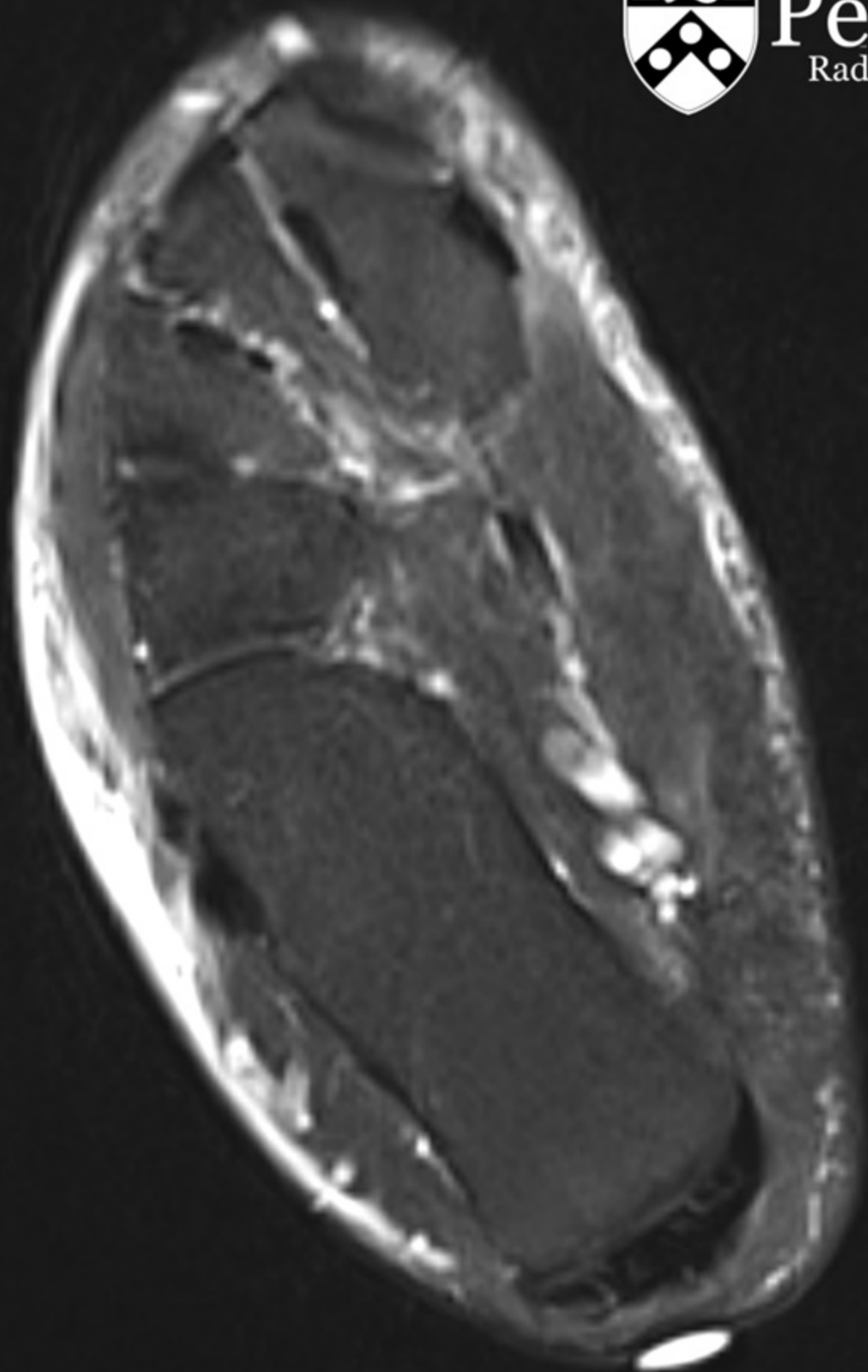
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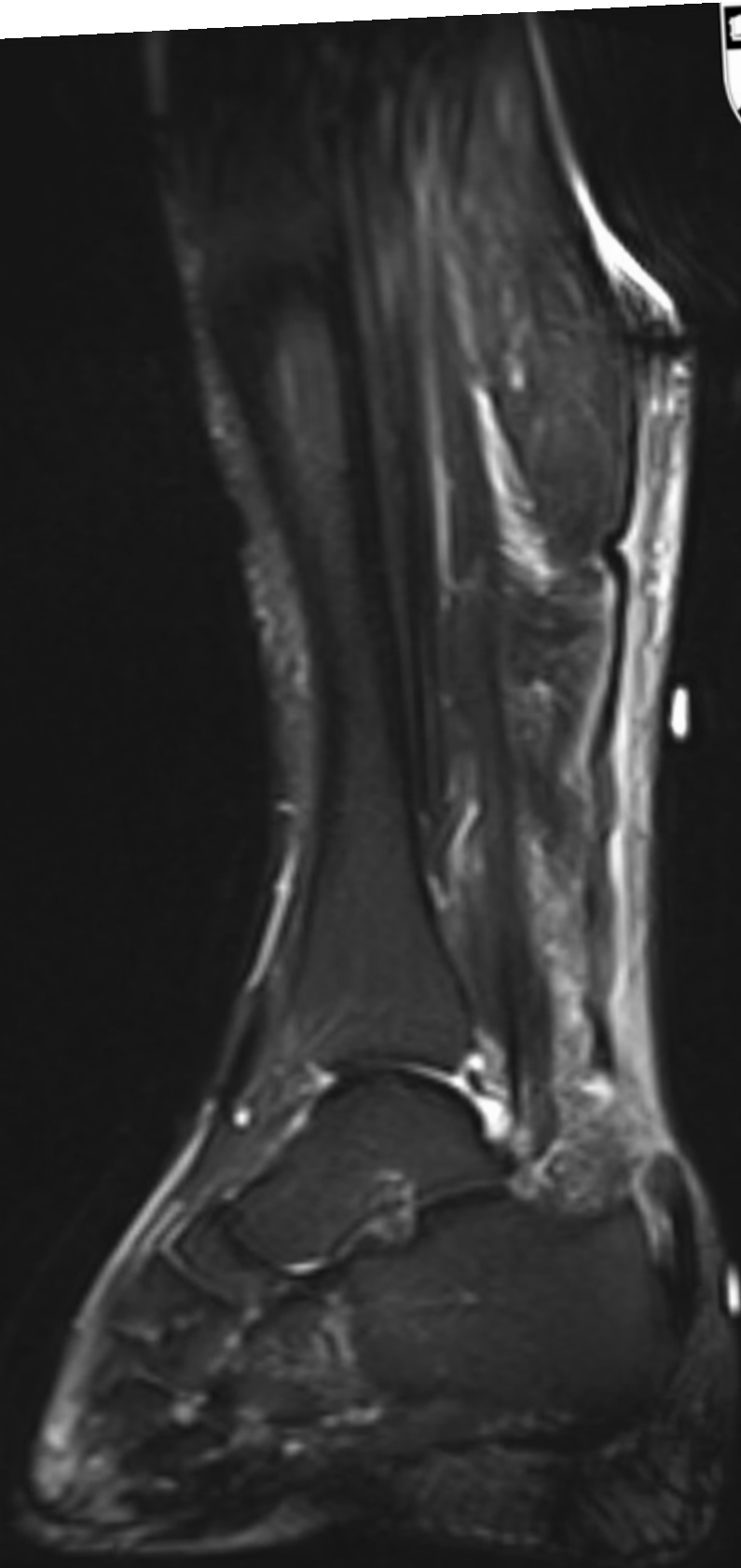
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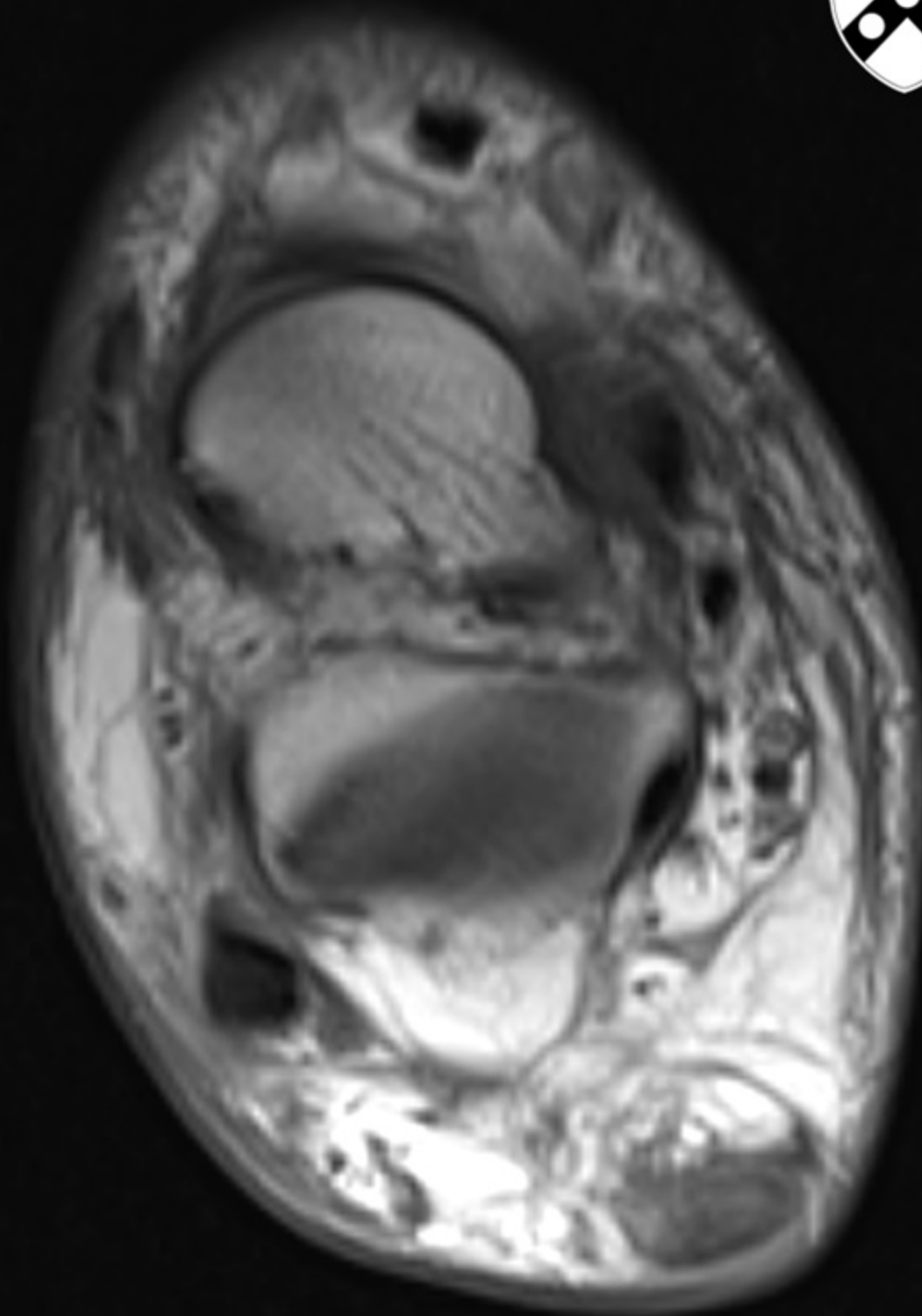


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## What is the diagnosis?

- ☐ Haglund syndrome
- ☐ Achilles tendon xanthoma
- ☐ Achilles tendon tear
- ☐ Achilles tendon ossification



## What is the diagnosis?

- ☐ Haglund syndrome
- ☐ Achilles tendon xanthoma
- ☒ Achilles tendon tear (correct!)
- ☐ Achilles tendon ossification



## Findings

There is proximal retraction and mild undulation of the Achilles tendon with discontinuity of the tendon and a gap measuring 2 cm located 3.5 cm from its insertion on the calcaneus, in keeping with complete Achilles tendon tear. Additionally, there is a suspected complete tear of the plantaris tendon. The peroneal, flexor digitorum longus, flexor hallucis longus, and extensor tendons are intact. There is mild associated edema of the soleus muscle. No acute osseous abnormality is identified.

## Differential diagnosis

- Complete tear of the Achilles tendon
- Haglund syndrome



of the plantaris tendon. The peroneal, flexor digitorum longus, flexor hallucis longus, and extensor tendons are intact. There is mild associated edema of the soleus muscle. No acute osseous abnormality is identified.

## Differential diagnosis

- Complete tear of the Achilles tendon
- Haglund syndrome
- Achilles tendon xanthoma
- Partial tear of the Achilles tendon
- Achilles tendon xanthoma
- Ossification of the Achilles tendon

**Diagnosis:** Complete (full-thickness) tear of the Achilles tendon

[VIEW YOUR SCORE](#)

## Additional questions

**Achilles tendon tears are more common in women.**

☐ True

☐ False

The question above accounts for 16% of your total score for this case.

**Which of the following has been associated with Achilles tendon tears?**

## Additional questions

**Achilles tendon tears are more common in women.**

☐ True

☒ False (correct!)

The question above accounts for 16% of your total score for this case.

**Which of the following has been associated with Achilles tendon tears?**

The question above accounts for 16% of your total score for this case.

**Which of the following has been associated with Achilles tendon tears?**

- ☐ Hypercholesterolemia
- ☒ Fluoroquinolone antibiotics
- ☐ Wearing high-heeled shoes
- ☐ Tarsal coalition



The question above accounts for 16% of your total score for this case.

**Which of the following has been associated with Achilles tendon tears?**

☐ Hypercholesterolemia

☒ Fluoroquinolone antibiotics (correct!)

☐ Wearing high-heeled shoes

☐ Tarsal coalition

The question above accounts for 16% of your total score for this case.

**The Achilles tendon is the only tendon in the body without a watershed zone.**

☐ True

☐ False

The question above accounts for 16% of your total score for this case.

**The Achilles tendon is made up of fibers arising from the gastrocnemius and soleus muscles.**

The question above accounts for 16% of your total score for this case.

**The Achilles tendon is the only tendon in the body without a watershed zone.**

☐ True

☒ False (correct!)

The question above accounts for 16% of your total score for this case.

**The Achilles tendon is made up of fibers arising from the gastrocnemius and soleus muscles.**

☒ False (correct!)

The question above accounts for 16% of your total score for this case.

**The Achilles tendon is made up of fibers arising from the gastrocnemius and soleus muscles.**

☐ True

☐ False

The question above accounts for 16% of your total score for this case.

**Blood supply to the Achilles tendon is from the anterior tibial artery.**



☐ False

The question above accounts for 16% of your total score for this case.

**The Achilles tendon is made up of fibers arising from the gastrocnemius and soleus muscles.**

☒ True (correct!)

☐ False

The question above accounts for 16% of your total score for this case.

**Blood supply to the Achilles tendon is from the anterior tibial artery.**

The question above accounts for 16% of your total score for this case.

**Blood supply to the Achilles tendon is from the anterior tibial artery.**

☐ True

☐ False

The question above accounts for 16% of your total score for this case.

[VIEW YOUR SCORE](#)

The question above accounts for 16% of your total score for this case.

**Blood supply to the Achilles tendon is from the anterior tibial artery.**

☐ True

☒ False (correct!)

The question above accounts for 16% of your total score for this case.

[VIEW YOUR SCORE](#)

# Achilles tendon tears

## Pathophysiology

- The Achilles tendon is made up of fibers arising from the gastrocnemius and soleus muscles.
- Blood supply to the tendon is from branches of the posterior tibial artery entering at the myotendinous junction and calcaneal insertion. The watershed zone 2 cm to 6 cm above the calcaneal insertion is most commonly affected, as sparse vascular supply increases the risk of a tear.
- An Achilles tendon tear is most commonly a sports-related injury (i.e., from squash, basketball, and tennis).
- Several pre-existing conditions increase the risk of a tear:
  - Tendinopathy
  - Fluoroquinolone treatment
  - Rheumatoid arthritis



- Rheumatoid arthritis
- Systemic conditions (i.e., diabetes mellitus, gout, connective tissue disorders)
- Steroid use
- Hyperparathyroidism
- Ochronosis
- Tears are classified using the Weinstabi classification system:
  - Type I: Inflammatory reaction
  - Type II: Degenerative changes
  - Type III: Partial tear
  - Type IV: Complete tear

## **Epidemiology**

- Achilles tendon tears are much more common in males.
- Occur commonly in the third to fifth decades of life.
- Incidence is 18 in 100,000.

- Achilles tendon tears are much more common in males.
- Occur commonly in the third to fifth decades of life.
- Incidence is 18 in 100,000.
- The Achilles is the most commonly injured tendon in the ankle.
- The left Achilles is torn more than the right.
- Typically, patients have no antecedent history of calf or heel pain prior to injury.

## **Clinical presentation**

- Patients often report a sudden “pop” sensation, a fall, and the inability to plantar flex the involved foot. Weak plantar flexion may be retained if the plantaris tendon is intact.
- A palpable tendon defect can sometimes be present on physical exam.
- The calf squeeze test is used to assess the integrity of the tendon. Circumferential compression of the thickest portion of the gastrocnemius muscle elicits passive ankle flexion if the Achilles tendon is intact.

# Imaging features

- Radiographs:
  - Loss of normal tendon definition
  - Fluid and soft-tissue swelling in the Kager fat pad
- MRI
  - Partial thickness and interstitial tears may show high signal on long repetition time (TR) sequences and tendon swelling greater than 7 mm anteroposteriorly.
  - Complete tear: A fluid-filled gap is seen in the tendon with retraction of the tendon ends.
- Ultrasound:
  - Ultrasound can distinguish complete from partial tears with 92% accuracy.
  - It can often detect if tendon ends approximate each other during plantar flexion, a finding which may aid in treatment planning.
  - With complete tears, a defect in the tendon and posterior acoustic shadowing are seen.



seen.

## Treatment

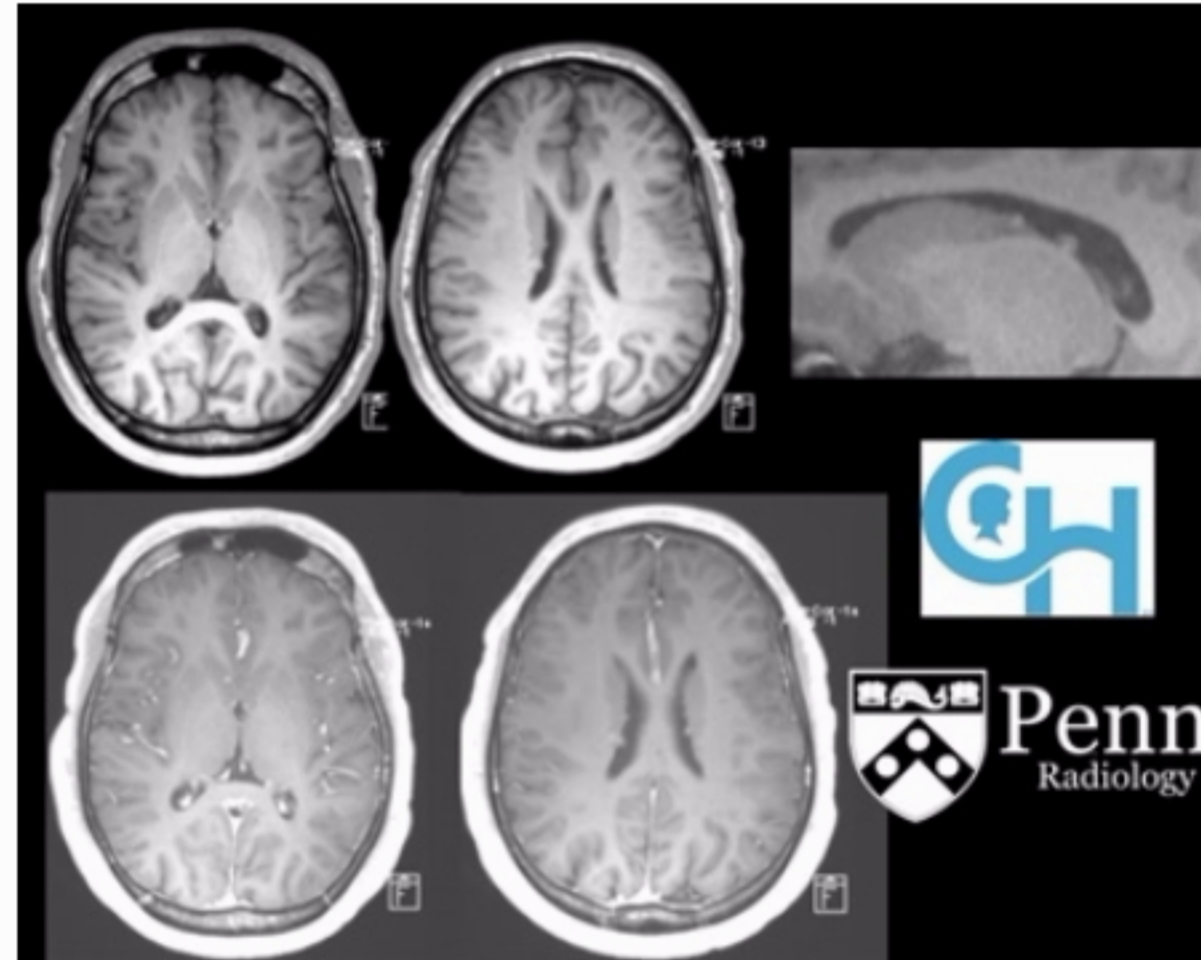
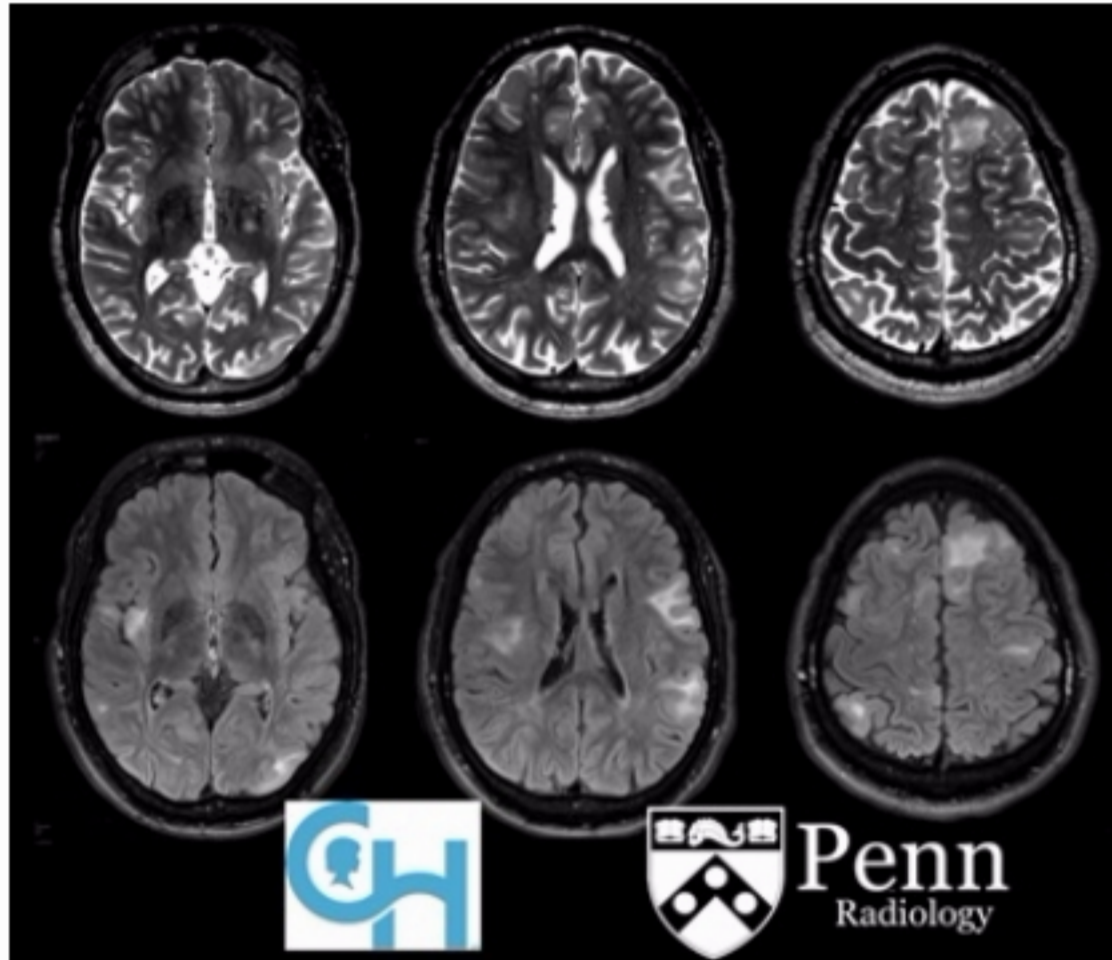
- Partial tears may initially be treated conservatively (i.e., cast immobilization), with subsequent surgery if conservative management fails.
- Complete tears are typically surgically repaired if the patient is a good surgical candidate.
  - Less than 3-cm diastasis: End-to-end anastomosis.
  - Greater than 3-cm diastasis: Typically requires tendon graft (usually from flexor hallucis longus).
- The nonsurgical rate of rerupture is 21%.
- The surgical rate of rerupture is 2% to 5%

## References

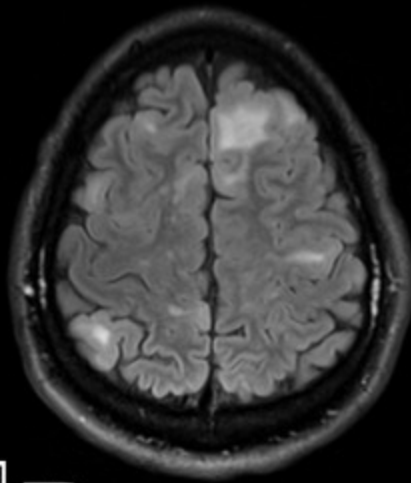
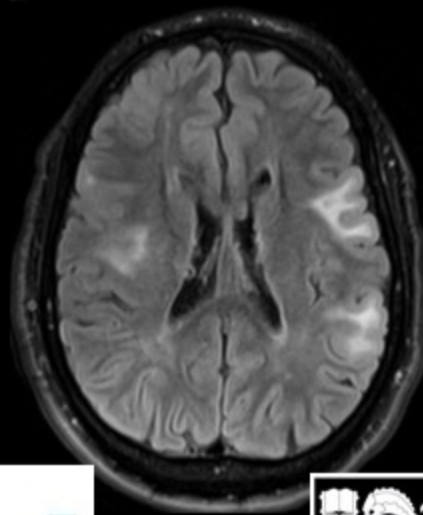
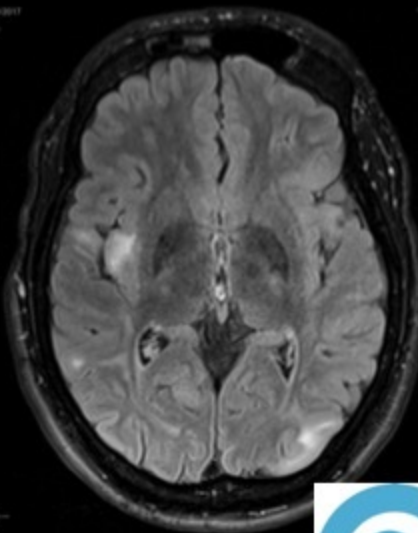
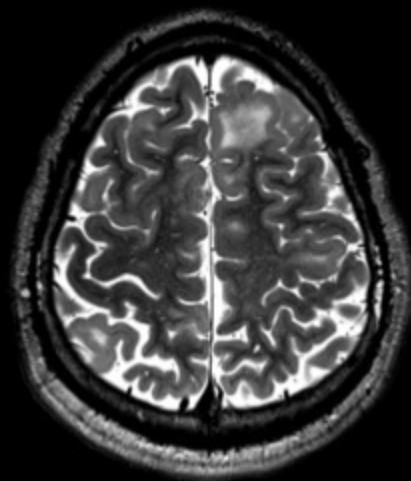
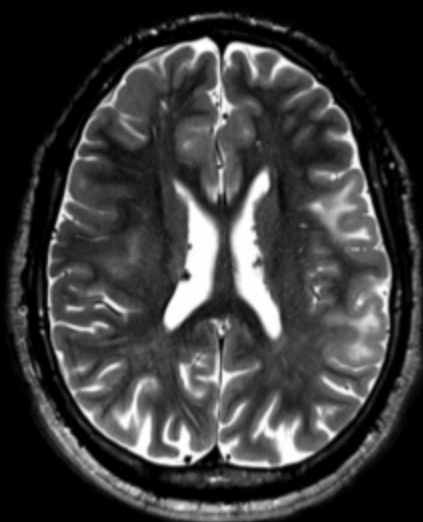
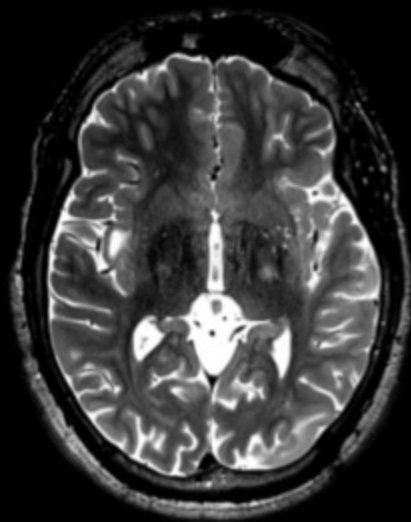
1. Galleis M, Connell DA. The Achilles tendon. *Semin Musculoskelet Radiol*.

**History:** A 17-year-old boy undergoing imaging workup for seizures and renal lesions.

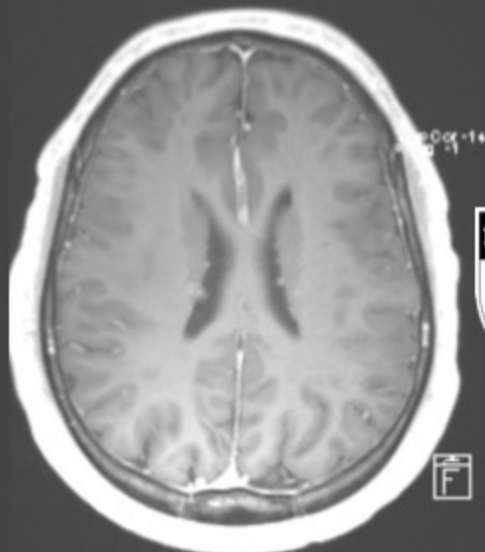
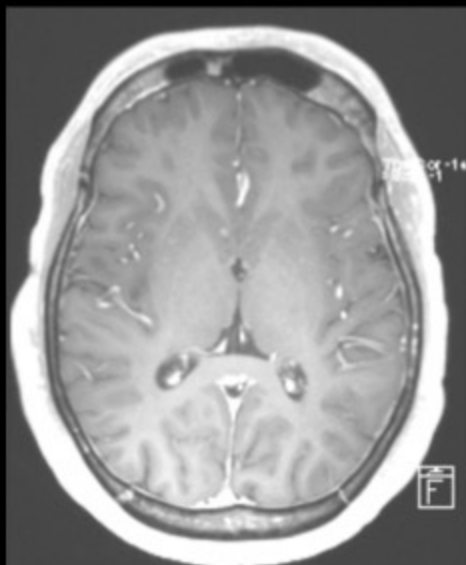
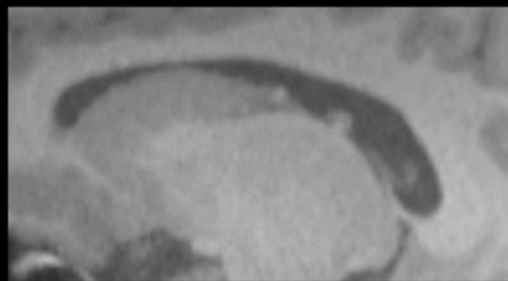
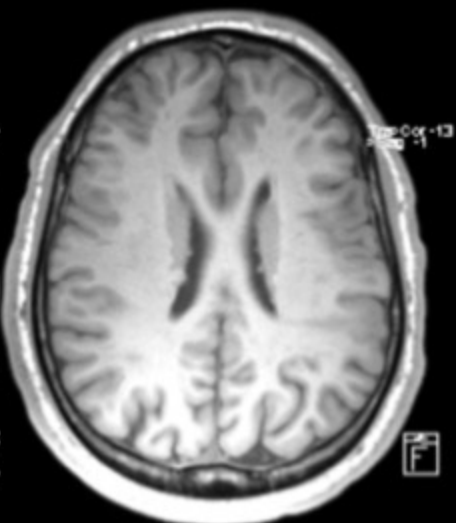
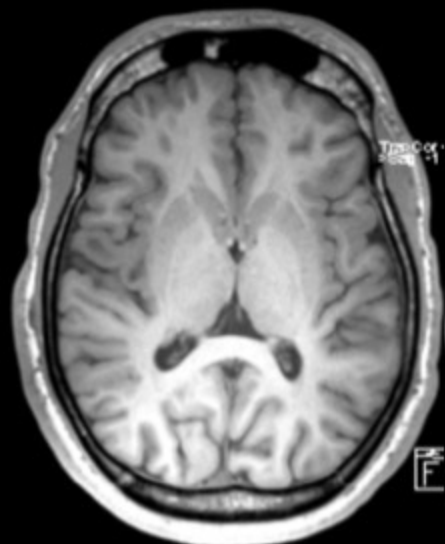
A brain MRI scan was performed. Axial T2-weighted, T2-weighted fluid-attenuated inversion-recovery (FLAIR), axial and sagittal T1-weighted, and T1-weighted postcontrast images are shown below. Click to enlarge.



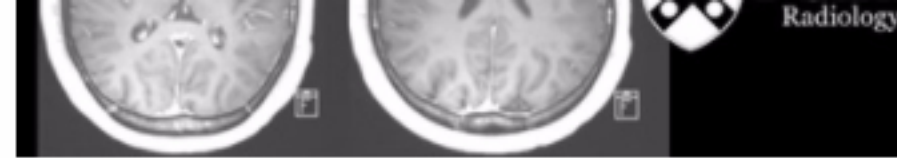




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**Where is there abnormal signal?**

☐ Frontal lobe

☐ Ventricular system

☐ Temporal lobe

☐ Parietal lobe

☐ All of the above

The question above accounts for 10% of your **total score** for this case.



**Where is there abnormal signal?**

- ☐ Frontal lobe
- ☐ Ventricular system
- ☐ Temporal lobe
- ☐ Parietal lobe

☒ All of the above (correct!)

The question above accounts for 10% of your **total score** for this case.

**There is abnormal enhancement.**

☐ True

☐ False

The question above accounts for 10% of your total score for this case.

**What is the most likely etiology of the lesions?**

☐ Vascular ischemic

☐ Autoimmune/vasculitis

☐ Infectious

☐ Neoplastic/genetic



**There is abnormal enhancement.**

☐ True

☒ False (correct!)

The question above accounts for 10% of your total score for this case.

**What is the most likely etiology of the lesions?**

☐ Vascular ischemic

☐ Autoimmune/vasculitis

☐ Infectious

☒ Neoplastic/genetic (correct!)

The question above accounts for 10% of your total score for this case.

**What is the most likely diagnosis?**

☐ Neurofibromatosis 1

☐ Neurofibromatosis 2

☐ Tuberous sclerosis

☐ Sturge-Weber syndrome

☐ Von Hippel-Lindau disease

The question above accounts for 10% of your total score for this case.

**Patients with this condition are at risk of developing**

The question above accounts for 10% of your total score for this case.

### What is the most likely diagnosis?

- ☐ Neurofibromatosis 1
- ☐ Neurofibromatosis 2
- ☒ Tuberous sclerosis (correct!)
- ☐ Sturge-Weber syndrome
- ☐ Von Hippel-Lindau disease

The question above accounts for 10% of your total score for this case.

**Patients with this condition are at risk of developing**

**Patients with this condition are at risk of developing hydrocephalus.**

☐ True

☐ False

The question above accounts for 10% of your total score for this case.

**The lesions seen protruding into the ventricles are called subcortical tubers.**

☐ True

☐ False

The question above accounts for 10% of your total score for this case.

**Patients with this condition are at risk of developing hydrocephalus.**

☒ True (correct!)

☐ False

The question above accounts for 10% of your total score for this case.

**The lesions seen protruding into the ventricles are called subcortical tubers.**

☐ True

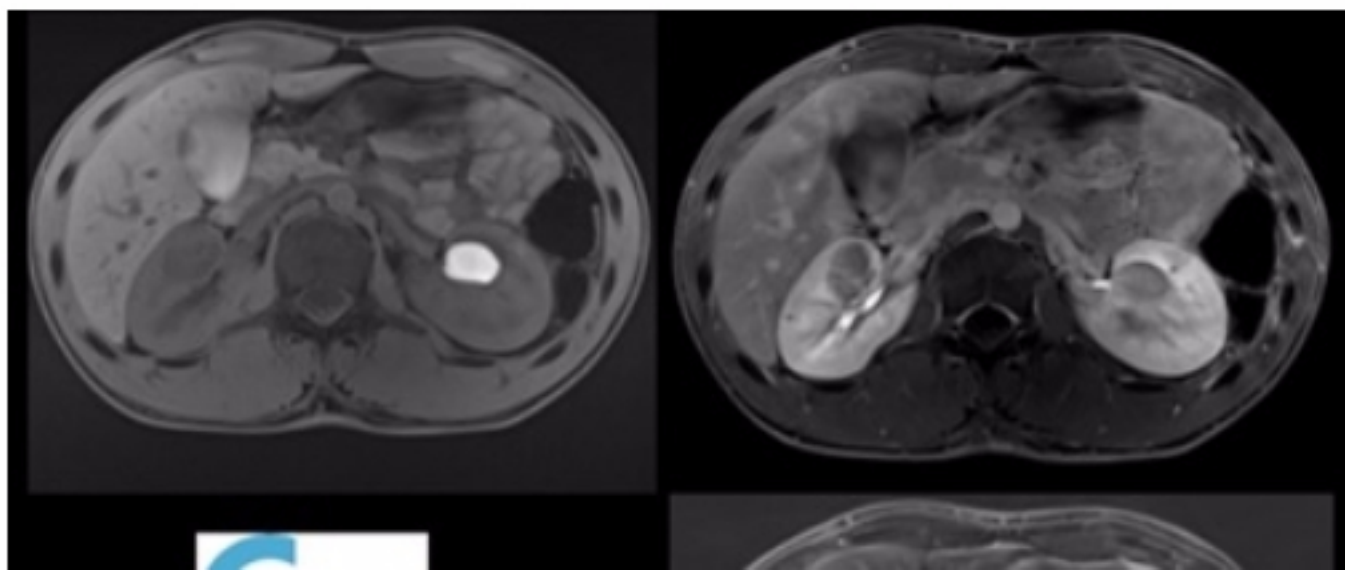
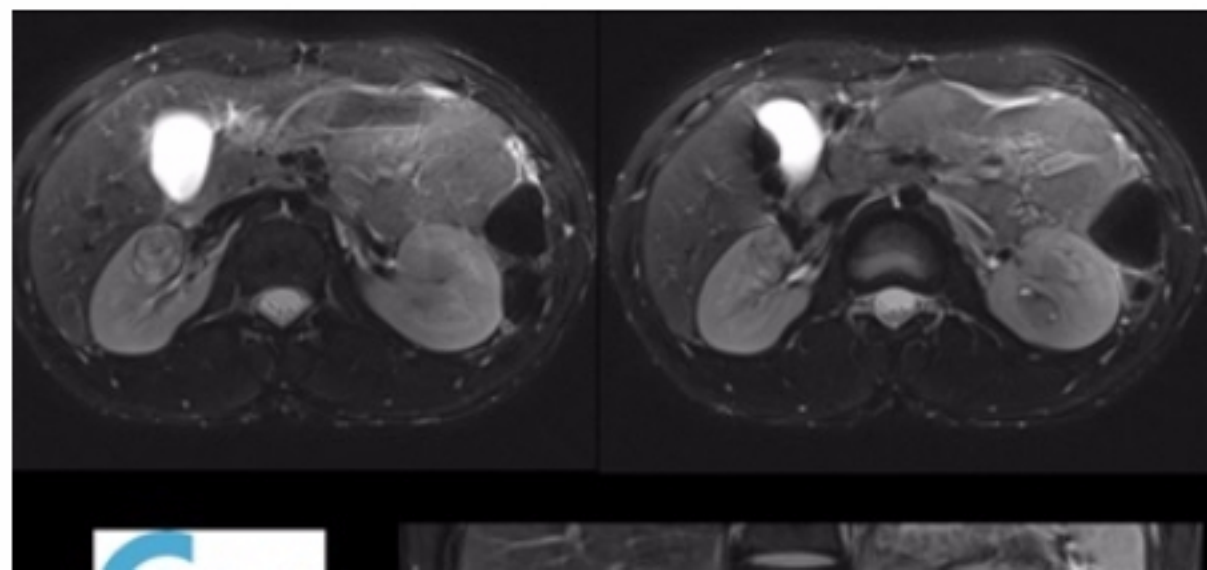
☒ False (correct!)

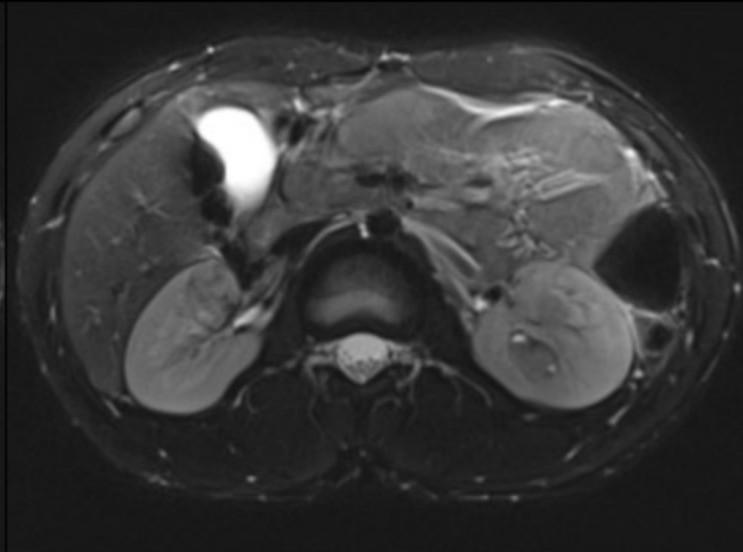
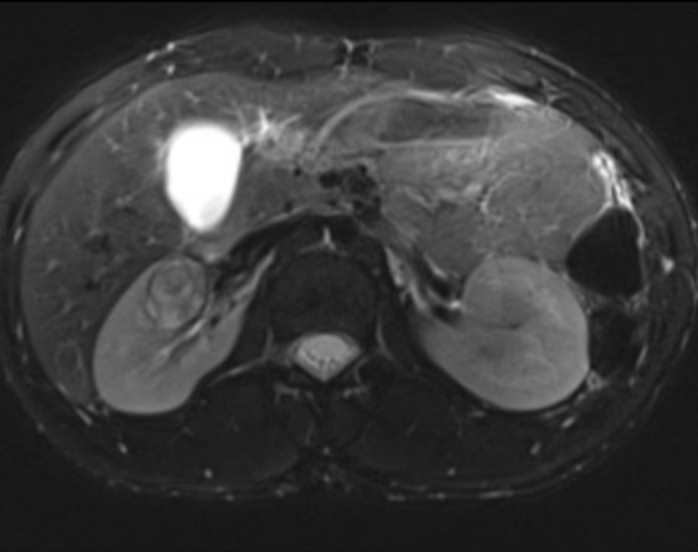
The question above accounts for 10% of your total score for this case.



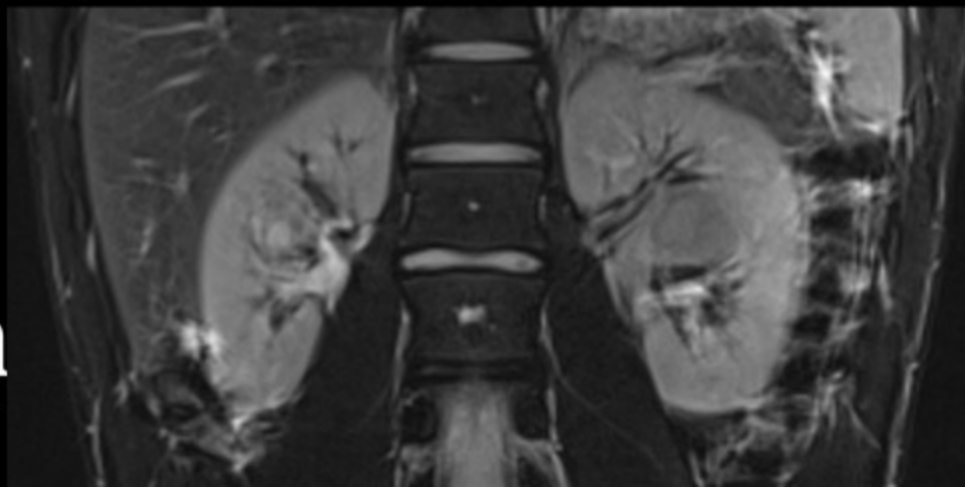
## Abdominal MR images

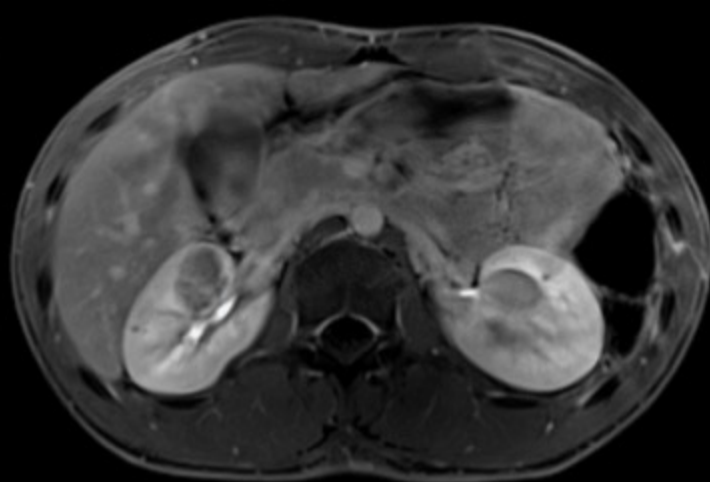
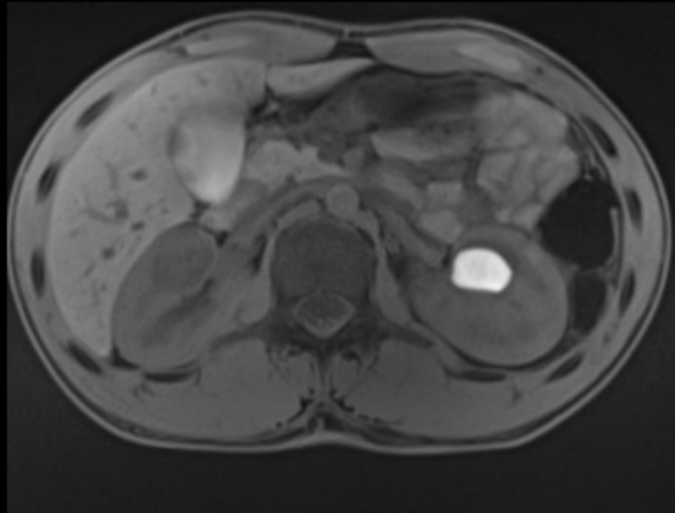
An MRI scan of the abdomen with and without contrast also was obtained the same day. Axial and coronal fat-saturated T2-weighted half-Fourier acquisition single-shot turbo spin-echo (HASTE) and axial T1-weighted fat-saturated volumetric interpolated breath-hold (VIBE), and fat-saturated precontrast, postcontrast, and subtraction images are shown below. Click to enlarge.



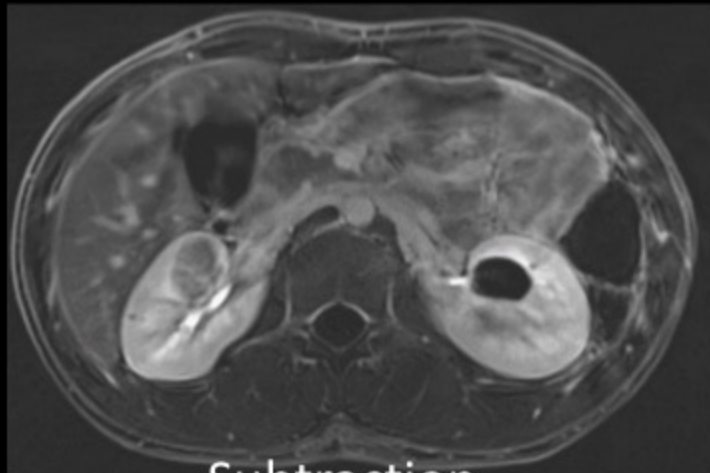


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Subtraction

**There are multiple renal lesions.**

☐ True

☐ False

The question above accounts for 10% of your total score for this case.

**There is enhancement.**

☐ True

☐ False

**There are multiple renal lesions.**

☒ True (correct!)

☐ False

The question above accounts for 10% of your total score for this case.

**There is enhancement.**

☒ True (correct!)

☐ False



☐ False

The question above accounts for 10% of your total score for this case.

**There is hemorrhage.**

☐ True

☐ False

The question above accounts for 10% of your total score for this case.

**What is the most likely diagnosis of the renal lesions?**

☐ False

The question above accounts for 10% of your total score for this case.

**There is hemorrhage.**

☒ True (correct!)

☐ False

The question above accounts for 10% of your total score for this case.

**What is the most likely diagnosis of the renal lesions?**

## What is the most likely diagnosis of the renal lesions?

☐ Hemorrhagic cysts

☐ Renal cell carcinomas

☐ Angiomyolipomas

☐ Liposarcomas

☐ Oncocytomas

## What is the most likely diagnosis of the renal lesions?

☐ Hemorrhagic cysts

☐ Renal cell carcinomas

☒ Angiomyolipomas (correct!)

☐ Liposarcomas

☐ Oncocytomas

## Findings

- **Brain MRI:** There are numerous foci of subcortical T2/FLAIR prolongation, many of which demonstrate radiating bands to the ventricular margins, consistent with cortical tubers. Multiple subependymal nodules are seen along the lateral ventricles and foramen of Monro. No abnormal enhancement or evidence of hydrocephalus is seen.
- **Abdominal MRI:** There are several well-circumscribed bilateral renal lesions measuring up to 2.6 cm. Signal within the lesions is somewhat heterogeneous, relatively hypointense on both T1- and T2-weighted images compared with the renal cortex. There is hemorrhage within one of the lesions on the left, and they display peripheral heterogeneous enhancement. **Although there is no macroscopic fat, these lesions are in keeping with angiomyolipomas given the history of tuberous sclerosis.**

## Differential diagnosis



peripheral heterogeneous enhancement. Although there is no macroscopic fat, these lesions are in keeping with angiomyolipomas given the history of tuberous sclerosis.

## Differential diagnosis

- Tuberous sclerosis
- Neurofibromatosis 1
- Neurofibromatosis 2
- Von Hippel-Lindau syndrome
- Birt-Hogg-Dubé syndrome
- Polycystic kidney disease
- Sturge-Weber disease

**Diagnosis:** Tuberous sclerosis

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# **Tuberous sclerosis (TS)**

## **Epidemiology and pathogenesis**

Tuberous sclerosis, also known as Bourneville disease, is a phakomatosis (neurocutaneous disorder) characterized by multiple tumors of the embryonic ectoderm (skin, eyes, and nervous system). It has an incidence of 1 in 6,000 to 12,000 and is associated with mutations in the TSC1 and TSC2 tumor suppressor genes. These genes encode the hamartin and tuberin proteins, which are involved in the regulation of cell growth and differentiation. The associated tumors are almost always benign hamartomas, although rarely they can become cancerous. The majority of cases (approximately 70%) are sporadic, while the remainder are inherited in an autosomal dominant fashion.

## **Clinical presentation**

The classic presentation consists of a clinical triad, seen in 30% of patients, which includes

## **Clinical presentation**

The classic presentation consists of a clinical triad, seen in 30% of patients, which includes seizures (75%), mental retardation (50%), and adenoma sebaceum (facial angiofibromas; 75%). Other common dermatologic presentations include ash leaf spots (hypopigmented spots), shagreen patches, and ungual fibromas. Patients sometimes first present with arrhythmias and heart failure in utero secondary to cardiac rhabdomyomas.

## **Diagnosis**

Genetic testing is usually diagnostic. The clinical presentation is also fairly pathognomonic. Imaging is crucial for understanding the exact malformations present in each individual and for presurgical planning.

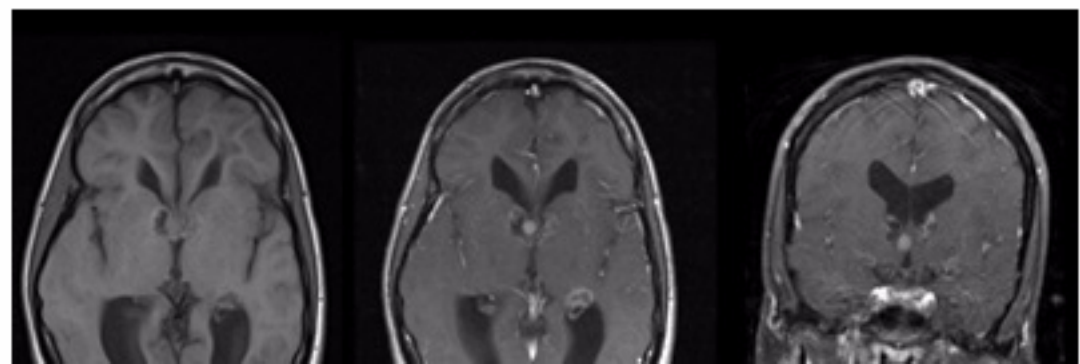
## **Imaging features**

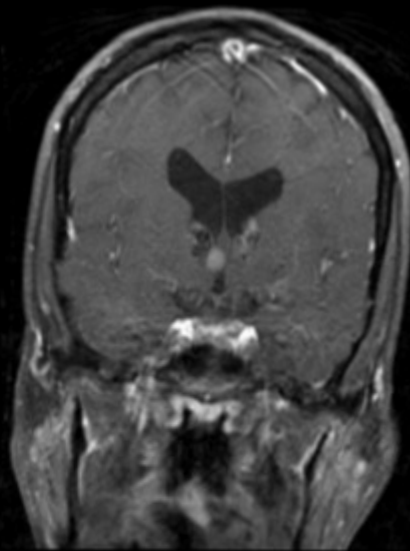
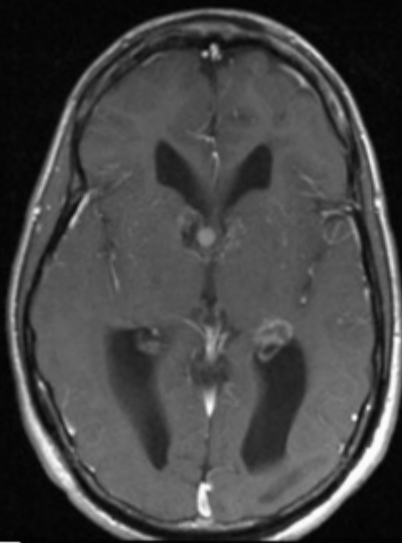
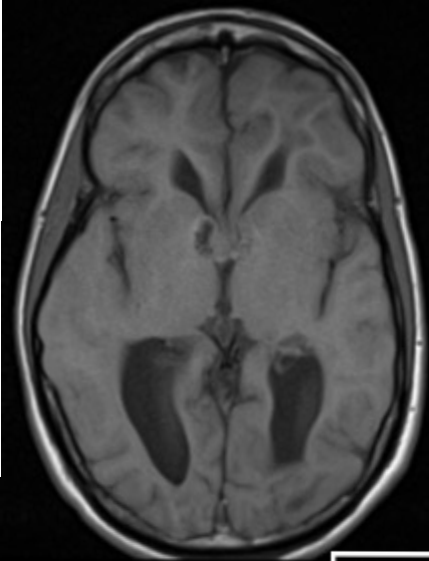


## Imaging features

- Neuroimaging findings: Cortical/subcortical tubers represent dysmorphic/disrupted neuronal layers.

Here are images from a sample case of a 38-year-old man with tuberous sclerosis demonstrating a 1-cm enhancing lesion in the region of the foramen of Monro with associated susceptibility artifact from calcifications that may represent a subependymal giant cell astrocytoma, noting that interval growth is would be the most important factor for a more definitive diagnosis.

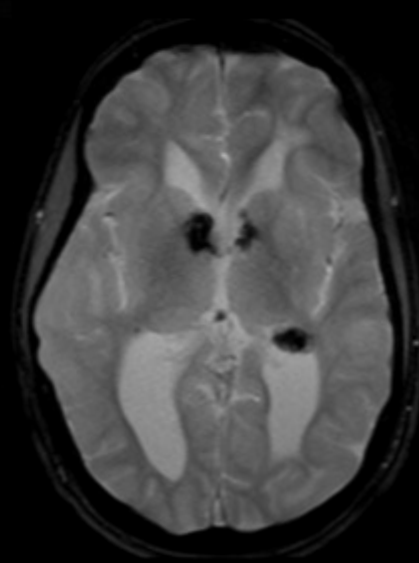
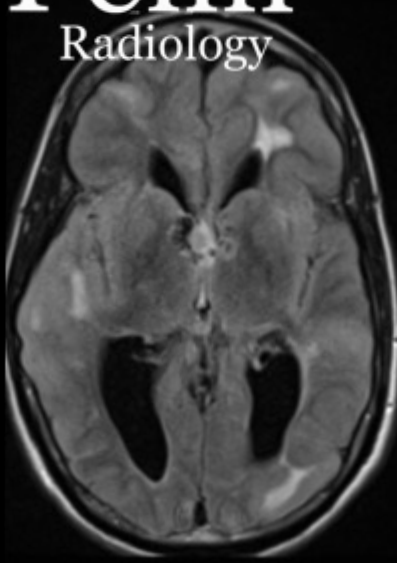
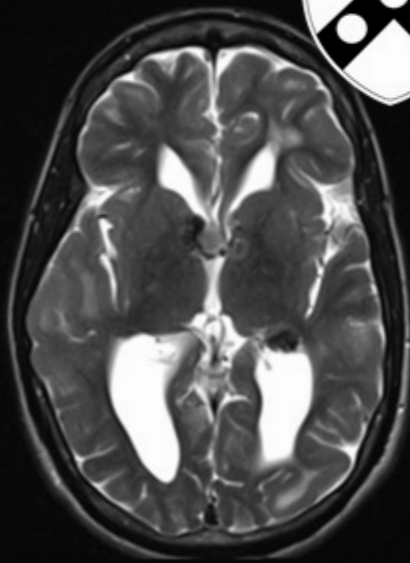




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- Renal imaging findings:
  - Angiomyolipomas (AMLs) are an abnormal collection of vessels, smooth muscle, and fat cells, seen in 55% to 75% of tuberous sclerosis patients.
  - Classically, AMLs can be distinguished from other enhancing renal lesions by macroscopic fat.
    - Often are hyperechoic on ultrasound.
    - Macroscopic fat seen on CT.
    - India ink etching artifact loss of signal seen on out-of-phase T1-weighted imaging.
    - However, they can often be lipid-poor.
  - AMLs in tuberous sclerosis tend to be bilateral and larger.
  - Size greater than approximately 4 cm or detection of aneurysms larger than 5 mm is associated with a higher risk of hemorrhage, necessitating pre-emptive treatment.
  - Additional renal manifestations include renal cysts, renal cell carcinoma, renal oncocytomas, and renal failure.

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- Size greater than approximately 4 cm or detection of aneurysms larger than 5 mm is associated with a higher risk of hemorrhage, necessitating pre-emptive treatment.
- Additional renal manifestations include renal cysts, renal cell carcinoma, renal oncocytomas, and renal failure.

## **Treatment and prognosis**

- Around 40% of patients will die from complications by age 35, which vary based on the patient's age.
- In utero and early in life, cardiac rhabdomyomas can cause heart failure and arrhythmias, but they often regress shortly before or after birth.
- Controlling seizures and supportive care for intellectual disability/autism are essential for treatment.
- Neurosurgical interventions may be required to treat hydrocephalus from



## Treatment and prognosis

- Around 40% of patients will die from complications by age 35, which vary based on the patient's age.
- In utero and early in life, cardiac rhabdomyomas can cause heart failure and arrhythmias, but they often regress shortly before or after birth.
- Controlling seizures and supportive care for intellectual disability/autism are essential for treatment.
- Neurosurgical interventions may be required to treat hydrocephalus from subependymal giant cell astrocytomas
- Surgical and interventional radiologic (often in combination) may be necessary to treat or prevent retroperitoneal hemorrhage from AMLs.
- Medications that inhibit the mTOR cell signaling pathway (i.e., everolimus) are used for certain indications.
- Later in life, pulmonary manifestations, including lymphangiomyomatosis, can lead to respiratory failure, and renal manifestations can lead to end-stage renal disease.

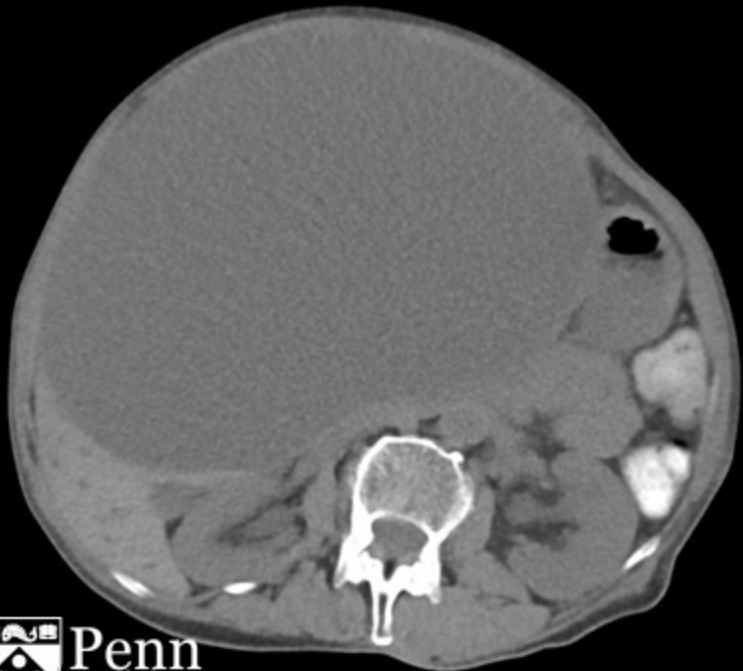
## History and CT images

Our appreciation is extended to Dr. Brian Currie, University of Pennsylvania Department of Radiology, for contributing this case.

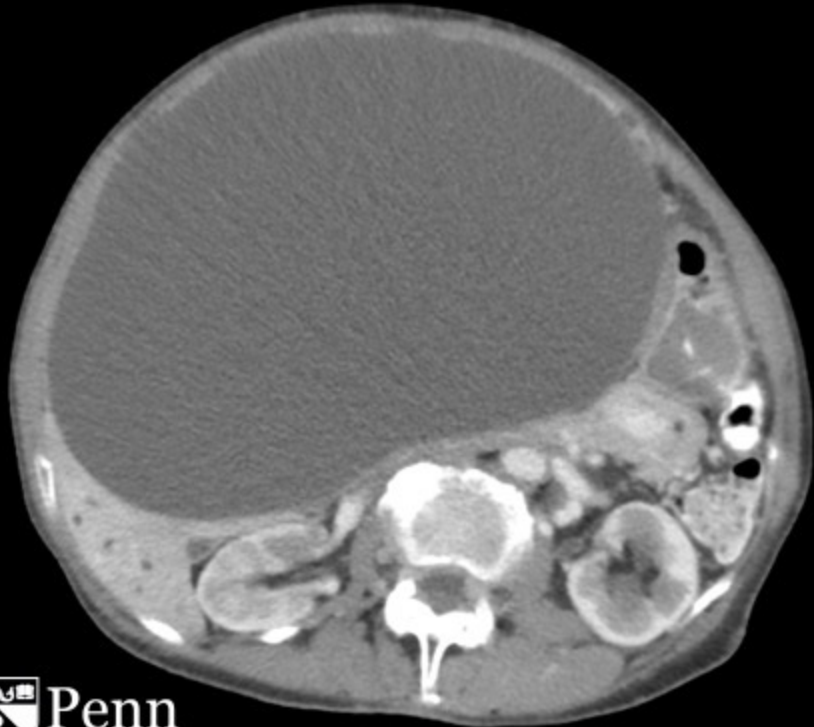
**History:** A 76-year-old woman presents to her primary care clinician for abdominal pain and bloating that has progressively worsened over the previous few months. After a physical exam reveals a distended and tense abdomen, the physician orders a CT scan of the abdomen and pelvis.

Unenhanced and contrast-enhanced CT images in soft-tissue window are shown below.  
Click to enlarge.





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**There is a lesion that most likely arises from which of the following organs?**

☐ Kidney

☐ Liver

☐ Adrenal gland

☐ Peritoneum

The question above accounts for 20% of your total score for this case.

**There is a lesion that most likely arises from which of the following organs?**

☐ Kidney

☒ Liver (correct!)

☐ Adrenal gland

☐ Peritoneum

The question above accounts for 20% of your total score for this case.



**Which of the following should NOT be included in the differential for this lesion?**

☐ Echinococcal cyst

☐ Simple liver cyst

☐ Hematoma

☐ Cystic hepatocellular carcinoma

☐ Biloma

Which of the following should NOT be included in the differential for this lesion?

☐ Echinococcal cyst

☐ Simple liver cyst

☐ Hematoma

☒ Cystic hepatocellular carcinoma (correct!)

☐ Biloma

## Findings

There is a large cystic lesion centered in the liver/biliary system, as determined by the “claw sign,” measuring 14.8 x 22.1 x 21.4 cm with mass effect on adjacent organs and intrahepatic biliary duct dilation. The mass contains thickened, nodular walls that exhibit mild enhancement.

## Differential diagnosis

- Echinococcal cyst
- Simple liver cyst
- Hematoma
- Biloma
- Abscess
- Biliary cystadenoma
- Biliary cystadenocarcinoma

There is a large cystic lesion centered in the liver/biliary system, as determined by the "claw sign," measuring 14.8 x 22.1 x 21.4 cm with mass effect on adjacent organs and intrahepatic biliary duct dilation. The mass contains thickened, nodular walls that exhibit mild enhancement.

## **Differential diagnosis**

- Echinococcal cyst
- Simple liver cyst
- Hematoma
- Biloma
- Abscess
- Biliary cystadenoma
- Biliary cystadenocarcinoma

**Diagnosis:** Biliary cystadenoma

**These lesions commonly arise from intrahepatic bile ducts.**

☐ True

☐ False

The question above accounts for 20% of your total score for this case.

**This lesion is more common in females.**

☐ True

☐ False



**These lesions commonly arise from intrahepatic bile ducts.**

☒ True (correct!)

☐ False

The question above accounts for 20% of your total score for this case.

**This lesion is more common in females.**

☒ True (correct!)

☐ False

The question above accounts for 20% of your total score for this case.

### **What is the best next step in management?**

☐ MRI with contrast

☐ Ultrasound

☐ Surgical resection

☐ Biopsy/drainage

The question above accounts for 20% of your total score for this case.

The question above accounts for 20% of your total score for this case.

### What is the best next step in management?

- ☐ MRI with contrast
- ☐ Ultrasound
- ☒ Surgical resection (correct!)
- ☐ Biopsy/drainage

The question above accounts for 20% of your total score for this case.

## Biliary cystadenomas

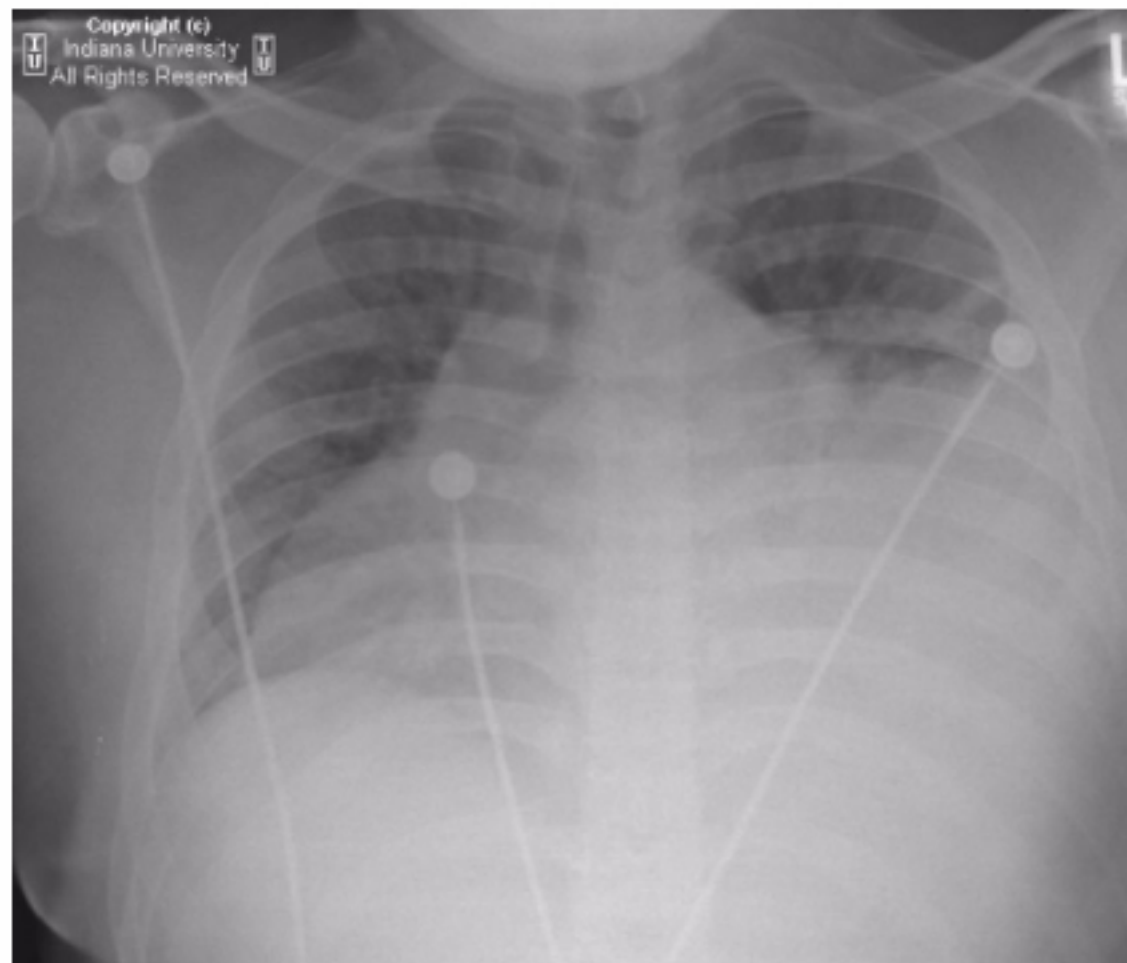
Biliary cystadenomas are rare hepatic tumors. They arise from the mucin-secreting bile duct epithelium and can grow quite large before exerting mass effect on adjacent structures and becoming symptomatic. There are two histologic subtypes: one containing ovarian stroma, which portends a better prognosis, and the other without ovarian stroma. Although biliary cystadenomas are benign, differentiation from a malignant biliary cystadenocarcinoma is quite difficult and, thus, detection usually results in resection.

The typical CT appearance of a biliary cystadenoma is a well-defined solitary uni- or multilocular cystic mass (with unilocular being far more prevalent) with a thick fibrous capsule with rare calcifications and also mural nodules. As the internal contents can vary from bilious, mucinous, hemorrhagic, or mixed, the MR appearance can be quite variable. Nonetheless, wall enhancement, nodularity, and a thickened fibrous capsule are features that differentiate a biliary cystadenoma from other benign hepatic cystic lesions.

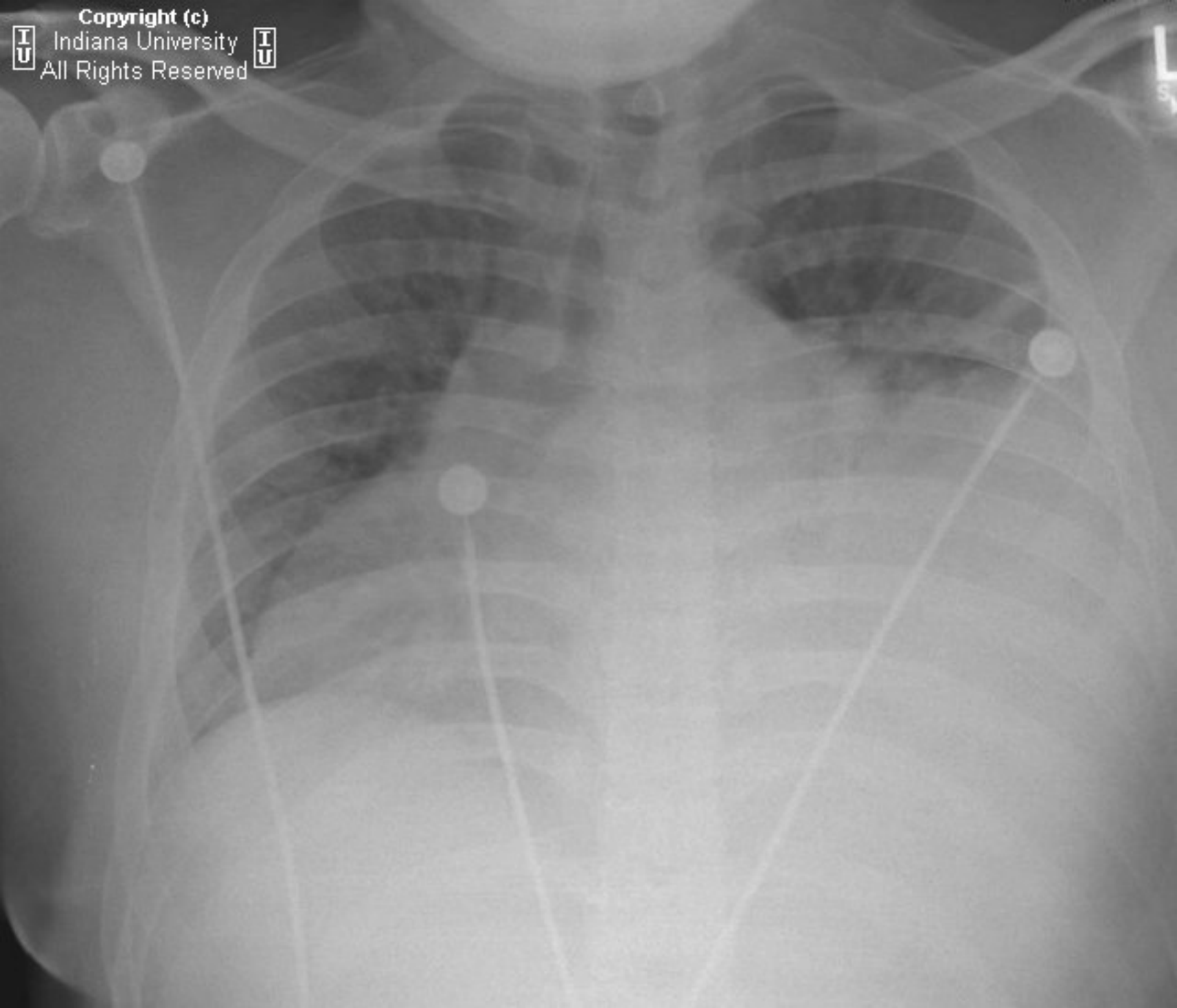


**History:** A man presents to the emergency room with syncope and facial lacerations.

Chest radiograph is shown below. Click to enlarge.









**Which choice best fits the radiographic findings?**

☐ Aortic transection

☐ Diaphragmatic hernia

☐ Mediastinal mass

☒ Pericardial effusion



**Which choice best fits the radiographic findings?**

☐ Aortic transection

☐ Diaphragmatic hernia

☒ Mediastinal mass (correct!)


☐ Pericardial effusion

☐ Aortic transecd

☐ Diaphragmati

☒ Mediastinal m

☐ Pericardial ef



There is mediastinal widening at level of carina with marked widening/lobulation of both the left and rights sides of the apparent heart border. The choice that best fits is mediastinal mass.

[**Explain this Answer**  
The question above

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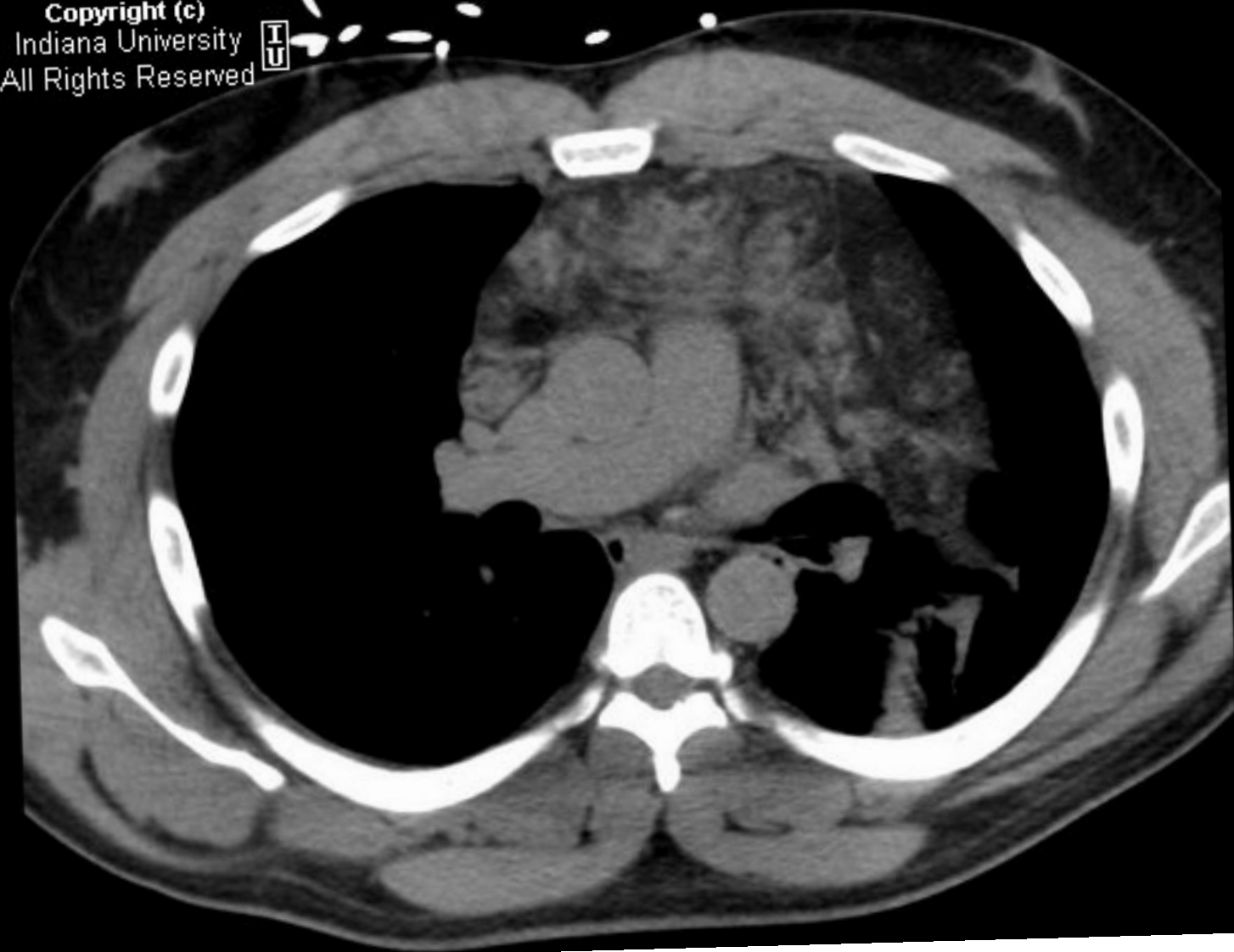
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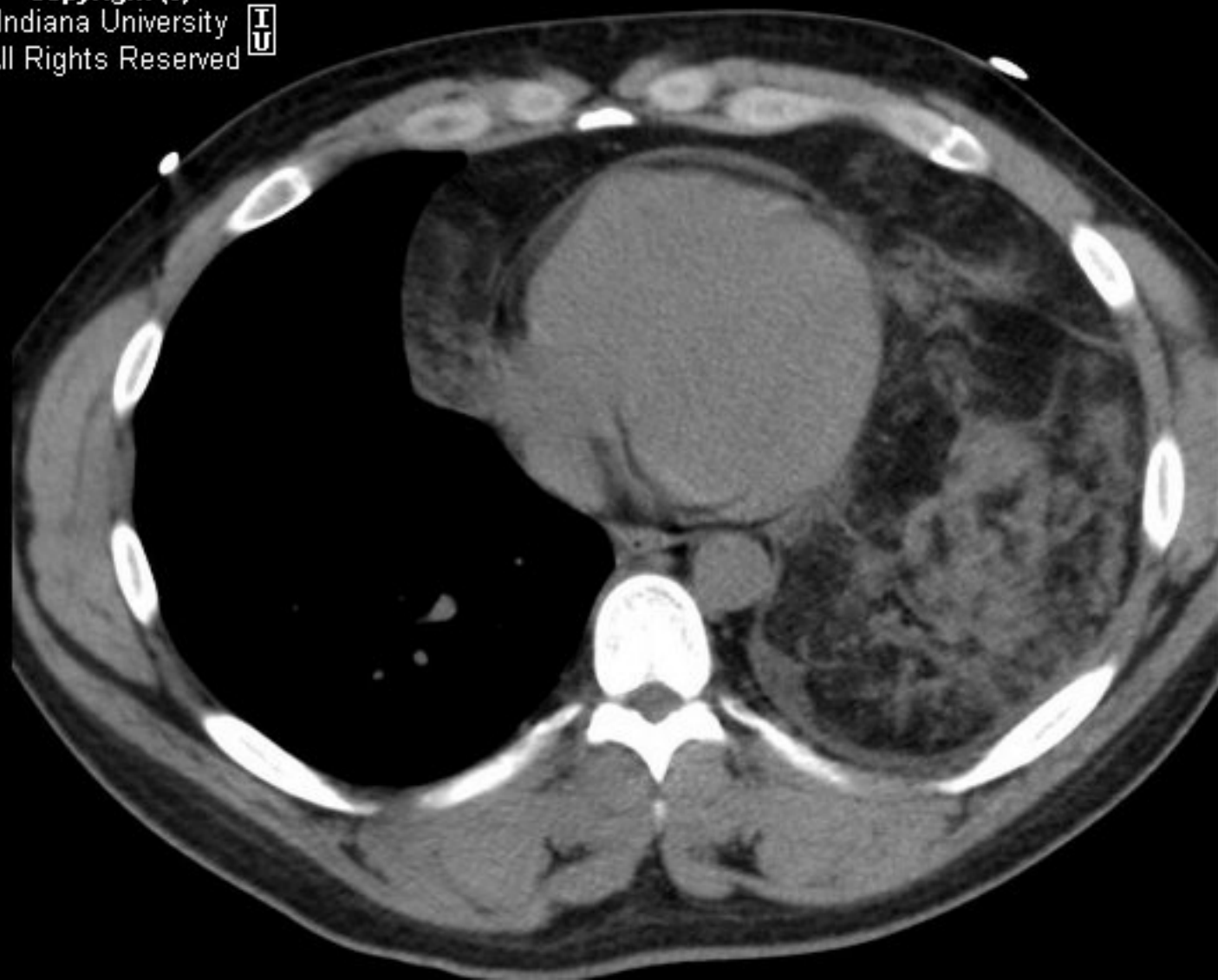


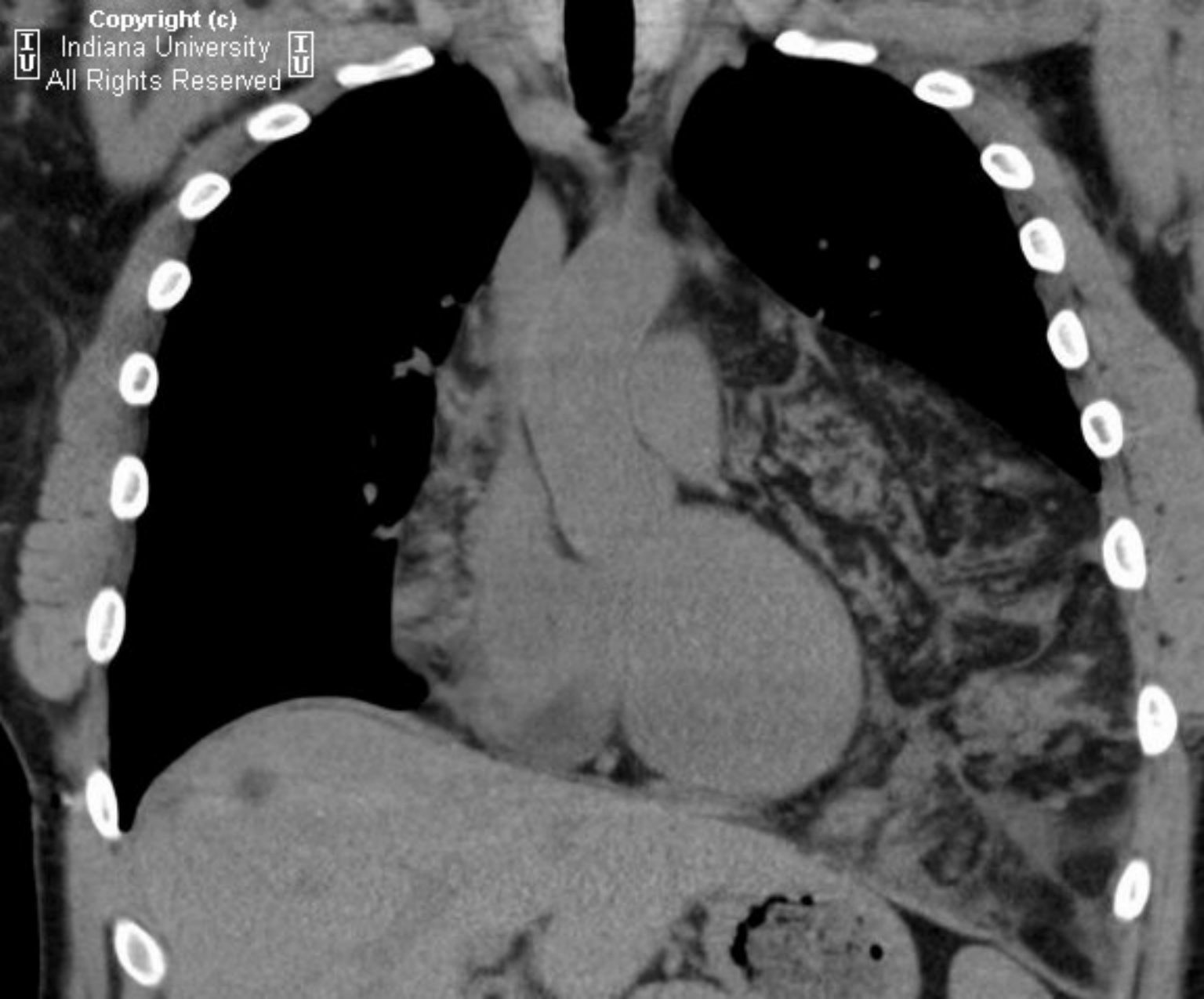


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## What is the diagnosis?

☐ Liposarcoma

☐ Mediastinal lipomatosis

☐ Thymolipoma

☐ Omental hernia

☐ Teratoma

## What is the diagnosis?

- ☐ Liposarcoma
- ☐ Mediastinal lipomatosis
- ☒ Thymolipoma (correct!)
- ☐ Omental hernia
- ☐ Teratoma



## **Findings**

There is a very large, fat-containing anterior mediastinal mass without evidence of significant mass effect. The mass is bulky but tends to conform to the shape of adjacent structures. The mass contains soft-tissue elements but no clearly cystic components or evidence of calcification. There is compressive atelectasis of the left lung base.

## **Differential diagnosis**

- Thymolipoma
- Liposarcoma
- Mediastinal lipomatosis/fat pad
- Omental herniation
- Mature teratoma (often cystic, with calcifications, and rarely conform to shape of adjacent structures)

**Diagnosis:** Thymolipoma

# Thymolipoma

- Thymolipoma is a rare, benign, slow-growing neoplasm composed of mature thymic tissue and adipose tissue.
- Accounts for 2% to 9% of all thymic neoplasms.
- Occurs most frequently in young adults, with mean age in mid-20s.
- In one study, 56% of patients were symptomatic.
- Thymolipoma is usually asymptomatic and manifests as a large anterior mediastinal mass; however, in one series, 56% of patients were symptomatic (upper respiratory infection, dyspnea, tachypnea, nonspecific chronic chest complaints).
- Histology: Composed of mature fat and thymic tissue. Some may have microscopic calcifications.
- It is usually detected incidentally at routine chest radiography and may occasionally mimic cardiomegaly or an elevated hemidiaphragm.
- Very rare association with myasthenia gravis. There have been reported associations with Grave's and aplastic anemia.
- Often very large before clinically evident. Mean size is 18 cm in longest dimension in



- Often very large before clinically evident. Mean size is 18 cm in longest dimension in one series.
- No malignant transformation.
- The anterior inferior mediastinum is the typical location.
- Fatty malleable consistency allows them to conform to the shape of adjacent structures, draping around heart and mediastinal structures.

## **Radiological overview and diagnosis**

- Radiography:
  - Mediastinal widening
  - Anterior mediastinal mass
  - Obscuration of heart border, diaphragm
- CT
  - Fat and soft tissue appear as linear strand and whorls of soft tissue embedded in fat or rounded islands of soft tissue surrounded by fat.

- CT
  - Fat and soft tissue appear as linear strand and whorls of soft tissue embedded in fat or rounded islands of soft tissue surrounded by fat.
  - Most have equal amounts of fat and soft tissue.
  - It is rare to see calcifications on CT.

## Key points

- Thymolipoma is a rare, benign, slow-growing neoplasm composed of mature thymic tissue and adipose tissue.
- CT findings are listed below:
  - Fat and soft tissue appear as linear strand and whorls of soft tissue embedded in fat or rounded islands of soft tissue surrounded by fat.
  - Most have equal amounts of fat and soft tissue.
  - It is rare to see calcifications on CT (helpful to distinguish from teratoma).

## History and MR images

Our appreciation is extended to Drs. Brian Currie and Ronnie Sebro, PhD, University of Pennsylvania Department of Radiology, for contributing this case.

**History:** A 31-year-old man presents with progressive left ankle pain for the past several months with an increase in the size of a palpable mass over the lateral ankle.

A radiograph was performed to evaluate the mass. The patient subsequently underwent further evaluation with an ankle MRI scan.

Unenhanced and enhanced T1-weighted images are shown below. Click to enlarge. In order: sagittal enhanced and unenhanced, coronal enhanced and unenhanced, and axial enhanced and unenhanced images.

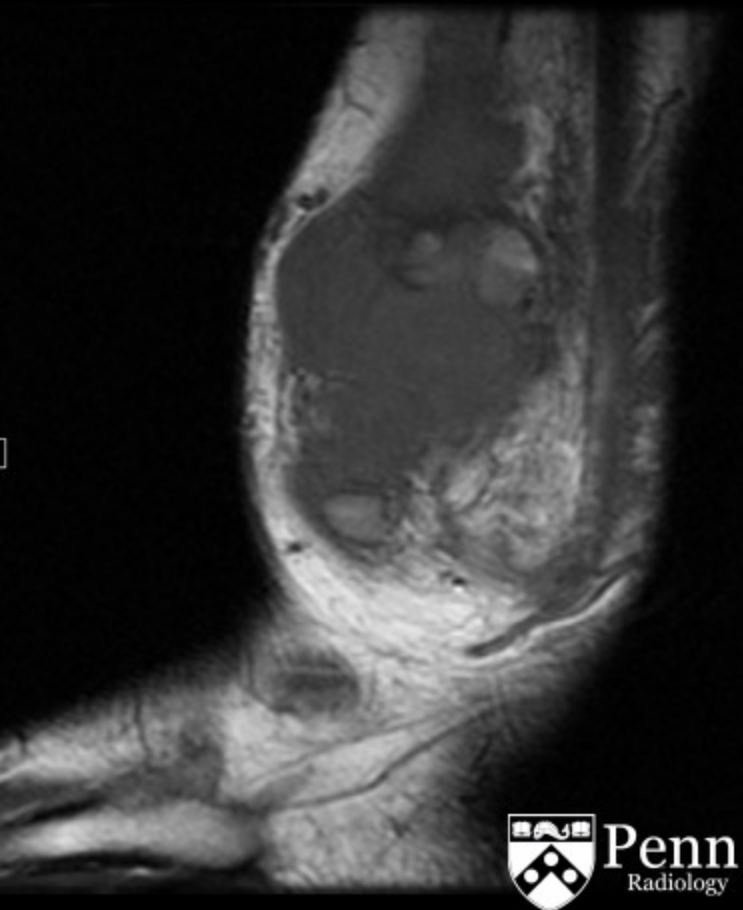




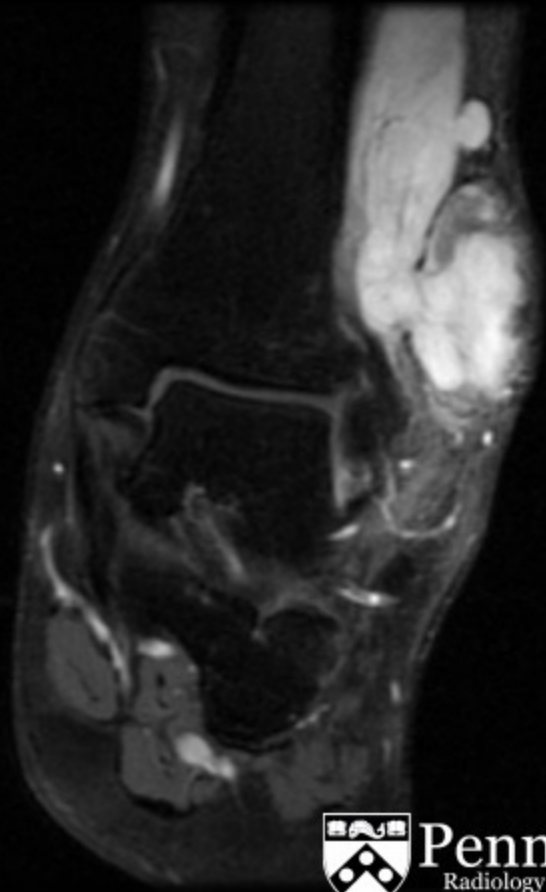
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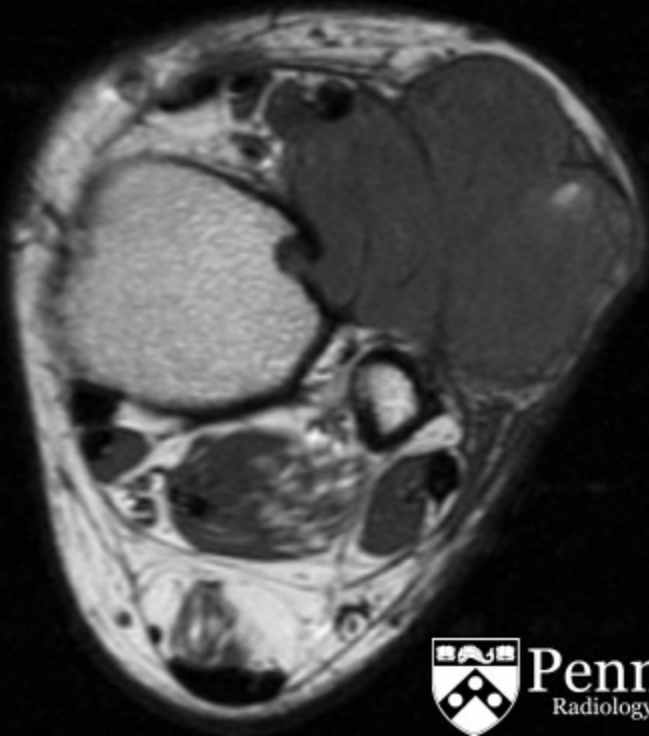
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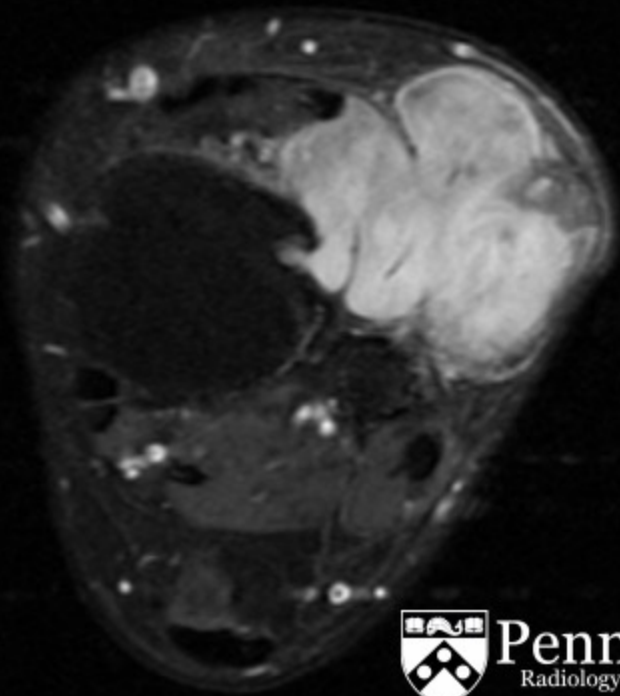


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Radiology

## What is the diagnosis?

☐ Chondromyxoid fibroma

☐ Fibrous dysplasia

☐ Adamantinoma

☐ Osteofibrous dysplasia

The question above accounts for 15% of your total score for this case.

## What is the diagnosis?

☐ Chondromyxoid fibroma

☐ Fibrous dysplasia

☒ Adamantinoma (correct!)

☐ Osteofibrous dysplasia

The question above accounts for 15% of your total score for this case.

## Additional questions

**This is a disease that commonly affects men in the second to third decades of life.**

☐ True

☐ False

The question above accounts for 15% of your total score for this case.

**Which of the following is the most common primary site for this lesion?**

## Additional questions

**This is a disease that commonly affects men in the second to third decades of life.**

☒ True (correct!)

☐ False

The question above accounts for 15% of your total score for this case.

**Which of the following is the most common primary site for this lesion?**



The question above accounts for 15% of your total score for this case.

**Which of the following is the most common primary site for this lesion?**

☐ Fibula

☐ Tibia

☐ Humerus

☐ Femur

The question above accounts for 15% of your total score for this case.

**Which of the following is the most common primary site for this lesion?**

☐ Fibula

☒ Tibia (correct!)

☐ Humerus

☐ Femur

**On plain radiographs, this lesion most closely resembles which of the following?**

☐ Enchondroma

☒ Osteofibrous dysplasia

☐ Osteoid osteoma

☐ Hemangioma

☐ Osteosarcoma

**On plain radiographs, this lesion most closely resembles which of the following?**

☐ Enchondroma

☒ Osteofibrous dysplasia (correct!)

☐ Osteoid osteoma

☐ Hemangioma

☐ Osteosarcoma

## **Findings**

There is a cortically based mass arising from the anterolateral distal tibial diaphysis with a large, lobulated, T1-isointense, T2-hyperintense, heterogeneously enhancing soft-tissue component.

## **Differential diagnosis**

- Chondromyxoid fibroma
- Fibrous dysplasia
- Adamantinoma
- Osteofibrous dysplasia

**Diagnosis:** Adamantinoma



## Additional questions

**This is a nonaggressive tumor with a low rate of local recurrence.**

☐ True

☐ False

The question above accounts for 14% of your total score for this case.

**This tumor almost never develops distant metastases**

## Additional questions

**This is a nonaggressive tumor with a low rate of local recurrence.**

☐ True

☒ False (correct!)

The question above accounts for 14% of your total score for this case.

**This tumor almost never develops distant metastases**

The question above accounts for 14% of your total score for this case.

**This tumor almost never develops distant metastases.**

☐ True

☐ False

The question above accounts for 14% of your total score for this case.

**Histologically, this tumor is comprised primarily of which of the following?**

☐ Epithelial cells that stain positive for cytokeratin

The question above accounts for 14% of your total score for this case.

**This tumor almost never develops distant metastases.**

☐ True

☒ False (correct!)

The question above accounts for 14% of your total score for this case.

**Histologically, this tumor is comprised primarily of which of the following?**

☐ Epithelial cells that stain positive for cytokeratin

☐ False (correct!)

The question above accounts for 14% of your total score for this case.

**Histologically, this tumor is comprised primarily of which of the following?**

☐ Epithelial cells that stain positive for cytokeratins

☐ Aberrant osteoclasts

☐ Disorganized sheets of lymphocytes



☒ False (correct!)

The question above accounts for 14% of your total score for this case.

**Histologically, this tumor is comprised primarily of which of the following?**

☒ Epithelial cells that stain positive for cytokeratins (correct!)

☐ Aberrant osteoclasts

☐ Disorganized sheets of lymphocytes

# Discussion

## Adamantinomas

Adamantinomas are rare ( $< 0.5\%$ ), indolent, malignant tumors that have a predilection for the tibia. Specifically, 80% to 85% of cases involve the tibia, and up to 97% involve the long bones, although involvement of almost all other major limb bones has been described. Additionally, there are very rare reports of solitary soft-tissue adamantinomas without bony involvement.

Patients typically present with a history of remote trauma (up to 60%) and also indolent pain, sometimes for many years, and with an occasionally palpable contour deformity or swelling in the involved limb.

## Imaging features

## Imaging features

Radiographic features usually consist of longitudinal, well-circumscribed, expansile, multilocular lytic lesion with sclerotic borders. **Adamantinomas** may be multifocal and take on a “soap bubble” appearance of multiple radiolucencies with ring-like sclerotic borders. When primarily in the tibia, they can even involve the ipsilateral fibula in rare instances.

MRI features of adamantinomas are somewhat nonspecific and consist of low/intermediate T1 and high T2 signal, with homogenous enhancement. There are, however, two main morphologic appearances: a solitary lobule or multifocal small nodules, which can both help in its differentiation from other malignant bone tumors. Nevertheless, the utility of MRI is somewhat limited and is, thus, more valuable for evaluation of the lesion's extent rather than its identity.

Histologically, adamantinomas are composed of epithelial cells that stain positive for cytokeratins on immunohistochemical studies, within an osteofibrous stroma. Due to this



Histologically, adamantinomas are composed of epithelial cells that stain positive for cytokeratins on immunohistochemical studies, within an osteofibrous stroma. Due to this appearance, it is theorized that adamantinomas arise from the displacement of skin epithelium during embryonic development, although its exact origin is still debated.

## **Treatment**

Adamantinomas are locally aggressive lesions, and their management consists of wide surgical excision. Metastatic disease occurs in 15% to 30% of patients and spreads both by the hematogenous and lymphatic routes, usually to the lungs, nearby lymph nodes, and adjacent bones. Radiation and chemotherapy have not proved effective in reducing local recurrence or metastatic disease, although there have been case reports of partial response with the chemotherapy drugs pazopanib and sunitinib.

## **References**

## History and CTA images

Our appreciation is extended to Dr. Erica Alexander, University of Pennsylvania Department of Radiology, for contributing this case.

**History:** A 39-year-old woman was transferred to a large tertiary center after an ultrasound scan revealed a deep venous thrombosis extending from the left internal jugular vein to the proximal left subclavian vein. She has a history of lupus, left-sided arteriovenous (AV) graft for dialysis, and multiple infections, including vertebral osteomyelitis.

Precontrast images from CT angiography (CTA) of the left upper extremity are shown below. Click to enlarge.

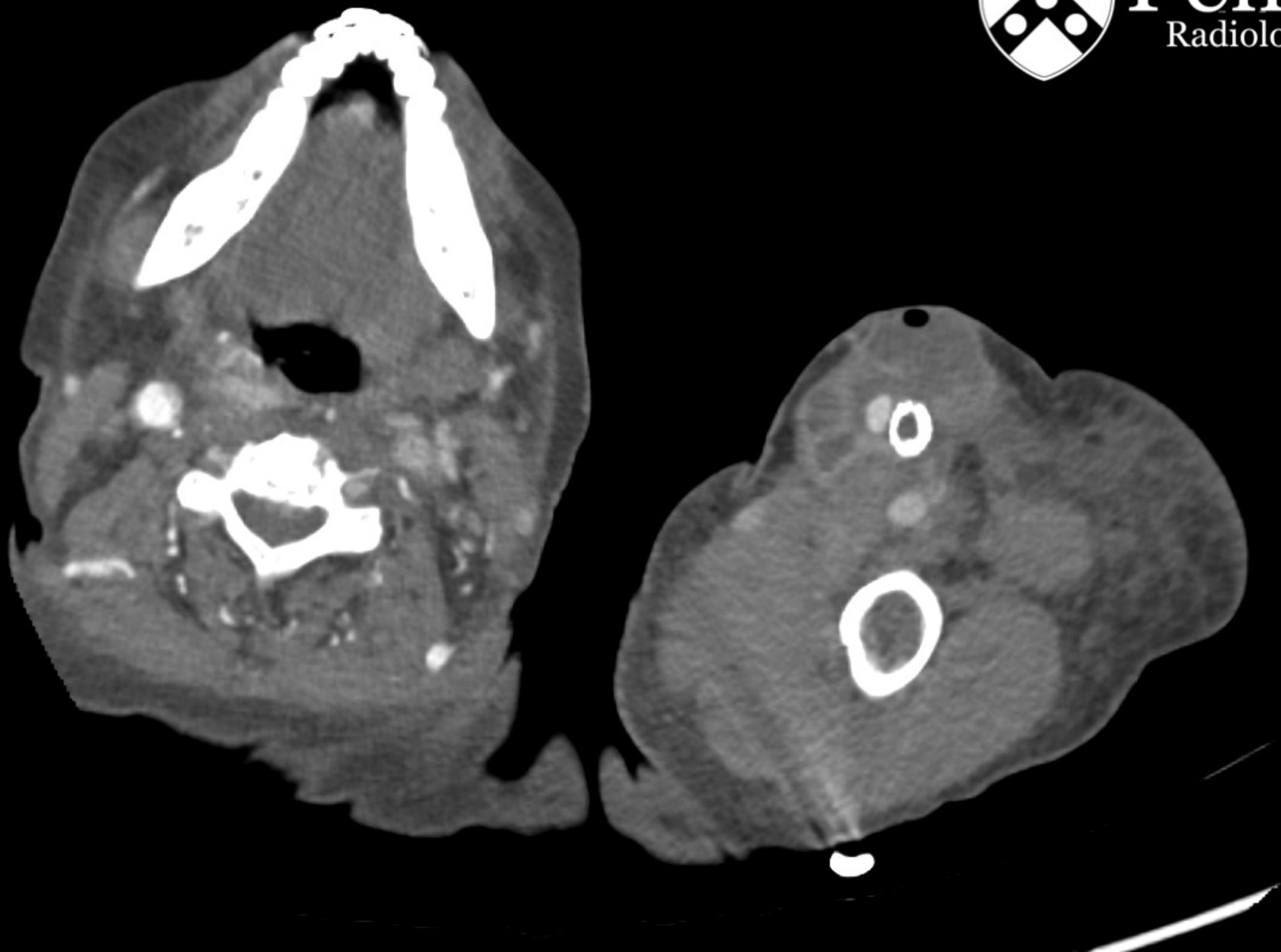






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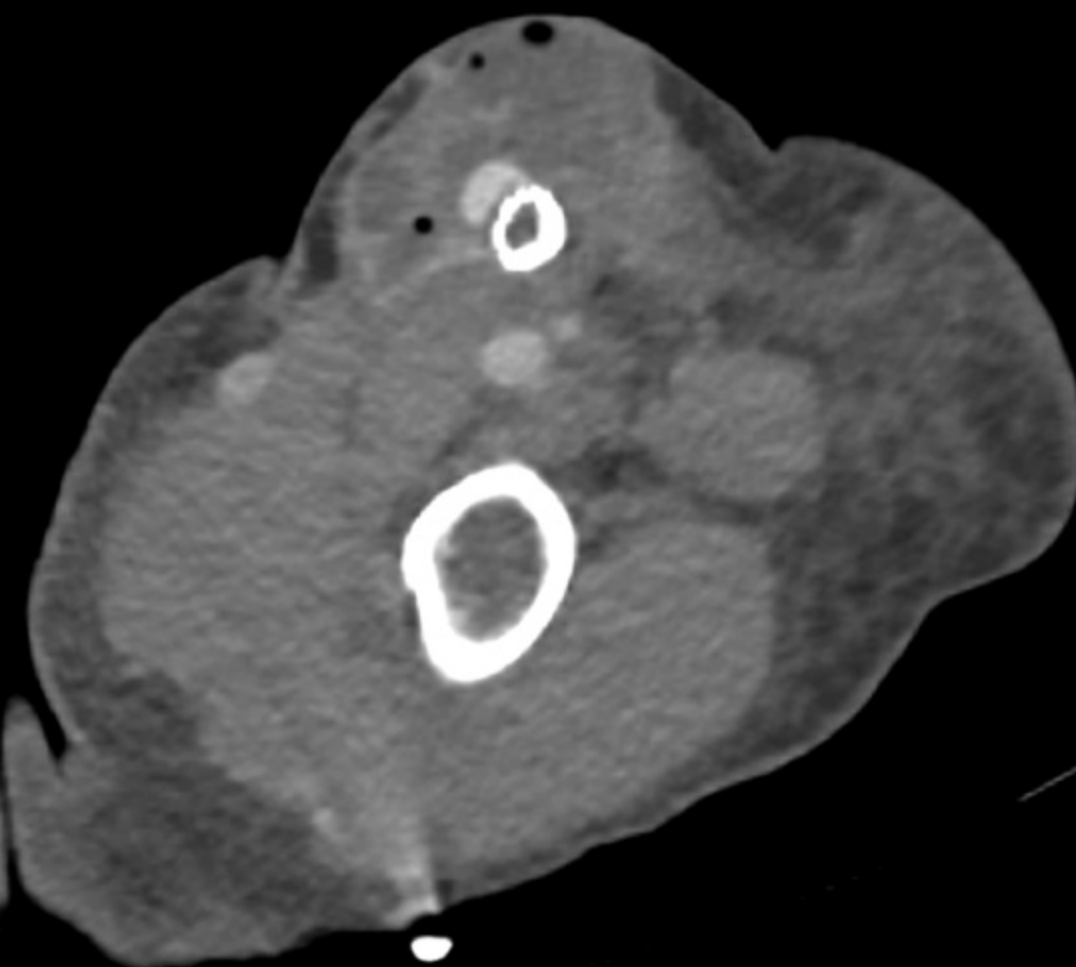
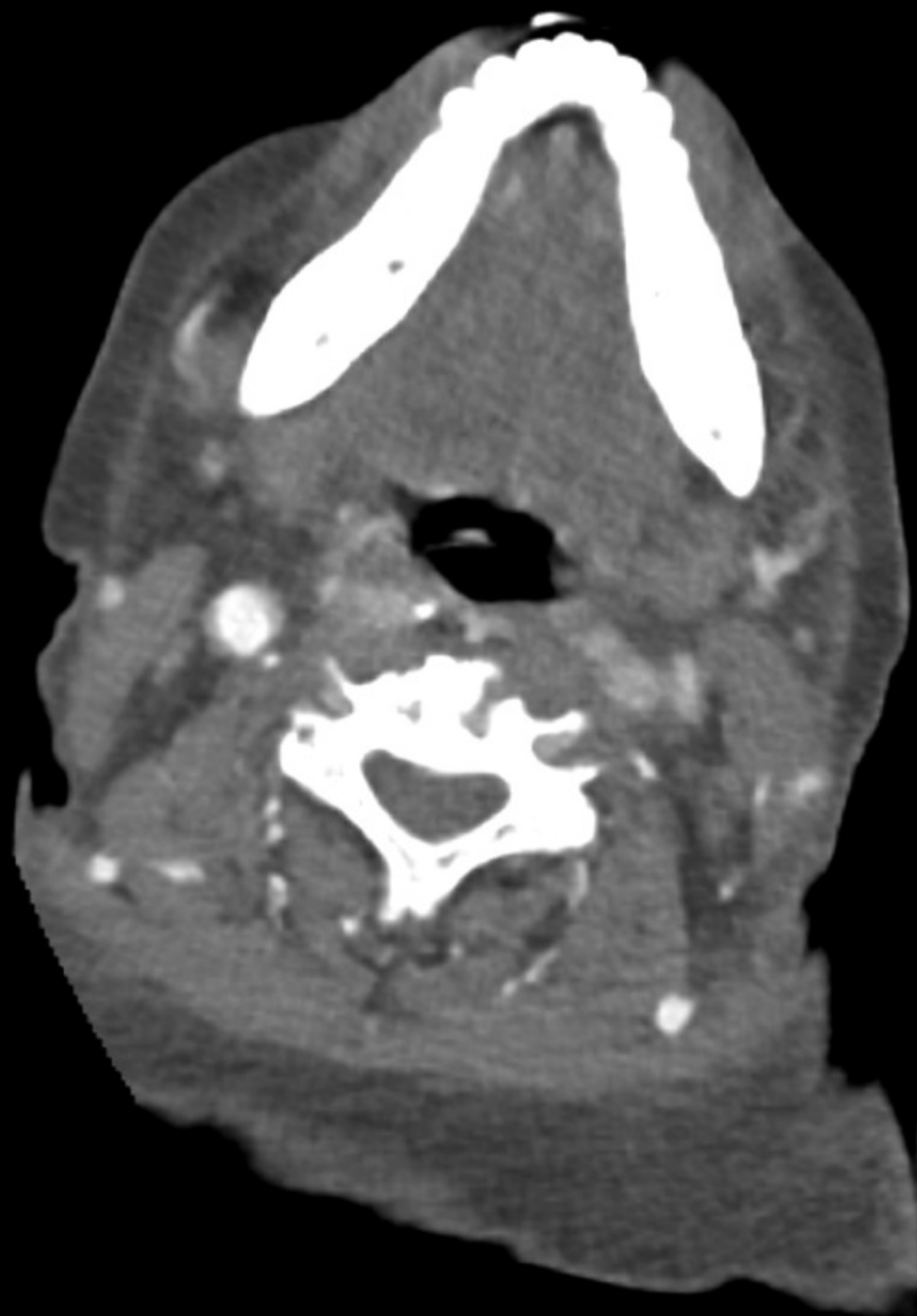
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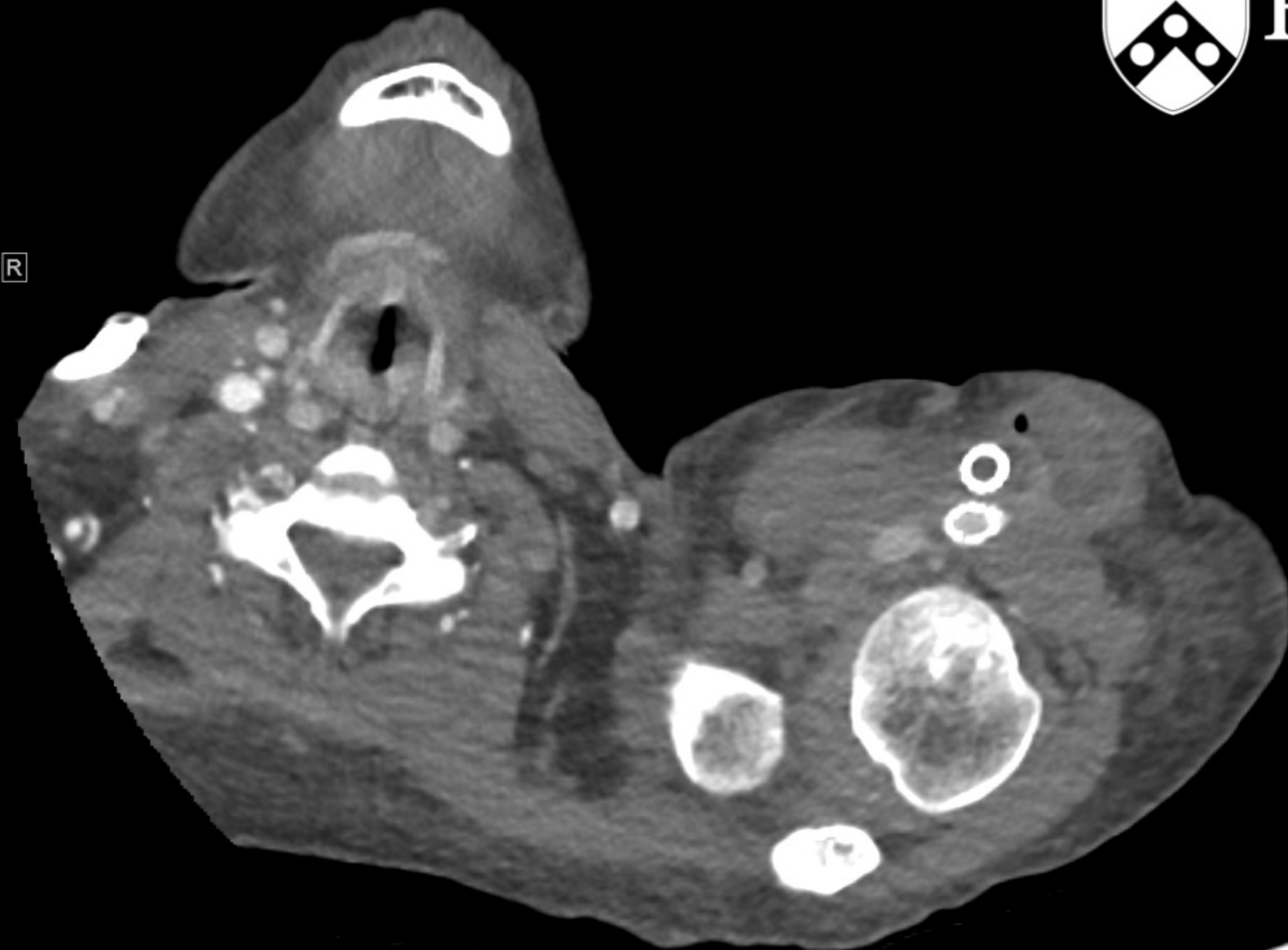
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## What is the abnormality?

☒ Thrombosis of the graft

☐ Stent fracture

☐ Malposition of the stent

☐ Abscess

The question above accounts for 20% of your total score for this case.

## What is the abnormality?

☐ Thrombosis of the graft

☐ Stent fracture

☐ Malposition of the stent

☒ Abscess (correct!)

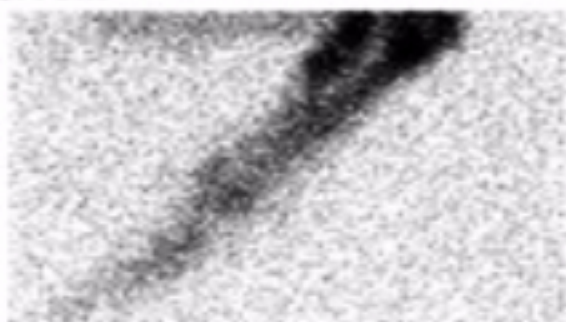
The question above accounts for 20% of your total score for this case.



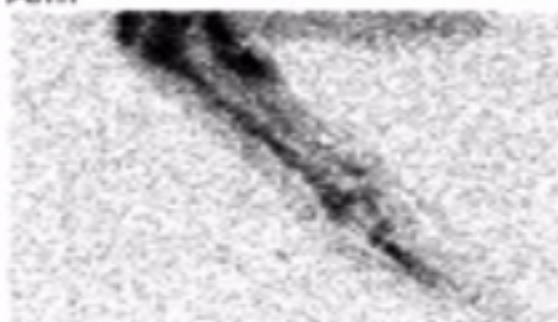
## WBC scan images

Autologous white blood cells (WBCs) labeled with a radioactive tracer were reinfused intravenously without incident. Then, 24 hours post injection, anterior and posterior planar scintigraphic images of the left and right upper arm were obtained. Click images below to enlarge.

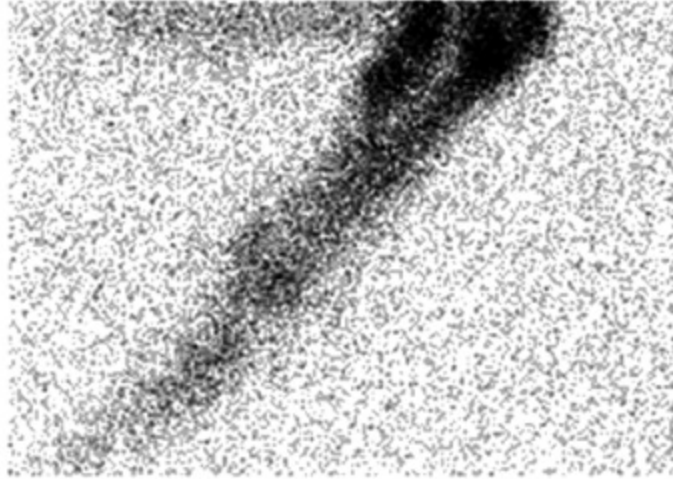
Right Upper Arm



Left Upper Arm

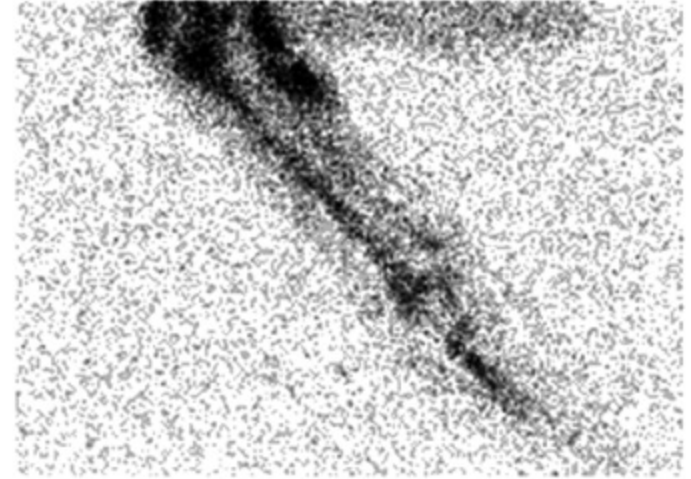


Right Upper Arm



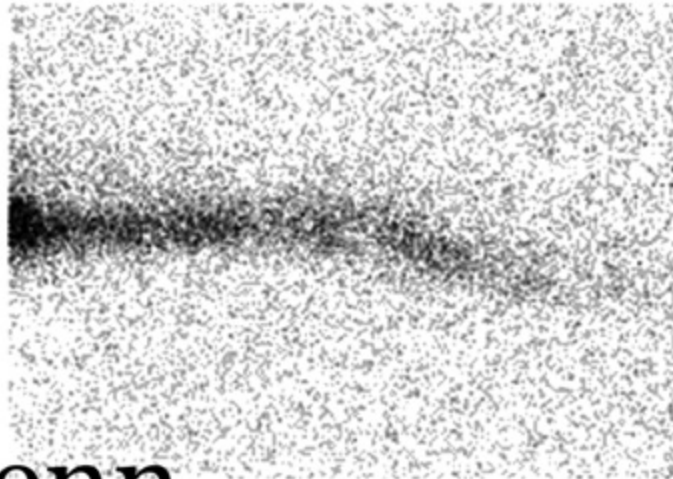
STATICS\_E  
12:11:55.0

Left Upper Arm

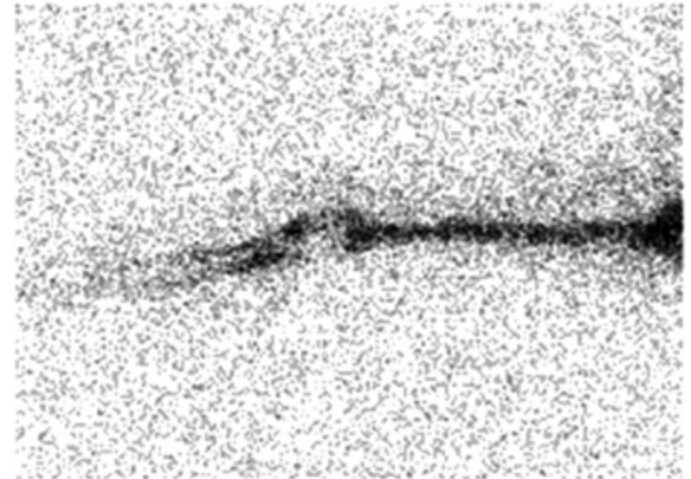


STATICS\_F  
12:11:55.0

Right Mid-Arm



Left Mid-Arm



RIGHT ARM\_F  
12:26:25.0

**Which radioactive tracer is used to label autologous white blood cells?**

☐ Indium-111

☐ Gallium-67

☐ Carbon-11

☐ Rubidium-82

The question above accounts for 20% of your total score for this case.

**Which radioactive tracer is used to label autologous white blood cells?**

☒ Indium-111 (correct!)

☐ Gallium-67

☐ Carbon-11

☐ Rubidium-82

The question above accounts for 20% of your total score for this case.

The question above accounts for 20% of your total score for this case.

**The radiotracer uptake in the bone marrow suggests an inflammatory/infectious state.**

☐ True

☐ False

The question above accounts for 15% of your total score for this case.

**Where is abnormal/increased radiotracer uptake seen in the above images?**

☐ Left lateral arm



☐ Rubidium-82

The question above accounts for 20% of your total score for this case.

**The radiotracer uptake in the bone marrow suggests an inflammatory/infectious state.**

☐ True

☒ False (correct!)

**[Explain this Answer]**

The question above accounts for 15% of your total score for this case.

☐ Carbon-11



X

☐ Rubidium-82

Labeled leukocytes can be taken up by normal bone marrow or in post-traumatic or postsurgical cases. False-positive results in the marrow-bearing skeleton can be avoided by obtaining a concomitant technetium-99m sulfur colloid study to evaluate the bones.

The question above

**The radiotracer inflammatory/infectious**

☐ True

☒ False (correct)

**Where is abnormal/increased radiotracer uptake seen in the above images?**

☐ Left lateral arm

☐ Left medial arm

☐ Right lateral arm

☐ Right medial arm

The question above accounts for 20% of your total score for this case.

**Rediabeled leukocyte uptake is not specific for infection and can occur in**

**Where is abnormal/increased radiotracer uptake seen in the above images?**

☐ Left lateral arm

☒ Left medial arm (correct!)

☐ Right lateral arm

☐ Right medial arm

The question above accounts for 20% of your total score for this case.

**Redicelabeled leukocyte uptake is not specific for infection and can occur in**

☐ Right medial arm

The question above accounts for 20% of your total score for this case.

**Radiolabeled leukocyte uptake is not specific for infection and can occur in any inflammatory process.**

☐ True

☐ False

The question above accounts for 15% of your total score for this case.

**Which of the following is an advantage of indium 111 labeled leukocytes as**



The question above accounts for 20% of your total score for this case.

**Radiolabeled leukocyte uptake is not specific for infection and can occur in any inflammatory process.**

☒ True (correct!)

☐ False

The question above accounts for 15% of your total score for this case.

**Which of the following is an advantage of indium-111-labeled leukocytes as compared to gallium-67 imaging?**

**Which of the following is an advantage of indium-111-labeled leukocytes as compared to gallium-67 imaging?**

- ☐ Better at imaging chronic infections
- ☐ Lower radiation dose
- ☐ Effective in severely leukopenic patients
- ☐ Greater concentration in abscessed tissues

The question above accounts for 10% of your total score for this case.

**Which of the following is an advantage of indium-111-labeled leukocytes as compared to gallium-67 imaging?**

- ☐ Better at imaging chronic infections
- ☐ Lower radiation dose
- ☐ Effective in severely leukopenic patients
- ☒ Greater concentration in abscessed tissues (correct!)

The question above accounts for 10% of your total score for this case.

## Findings

- **CTA:** There is a dialysis fistula in the left upper extremity. A fluid collection surrounds the arterial limb, and there is air in the collection at the level of the proximal humerus. This collection connects to a multiloculated air-containing collection that extends to the skin more anteriorly.
- **Tagged WBC study:** There is a focal area of increased radiotracer activity in the soft tissues along the medial aspect of the left upper arm, at the site of the known AV graft. The most intense activity correlates with the location of the collection surrounding the AV graft seen on CTA. There is also symmetric radiotracer uptake in the bilateral humeri and proximal radius and ulna, in keeping with physiologic distribution.

## Differential diagnosis -- increased radiotracer uptake in a WBC scan

- Uninfected postsurgical wound



AV graft seen on CTA. There is also symmetric radiotracer uptake in the bilateral humeri and proximal radius and ulna, in keeping with physiologic distribution.

### **Differential diagnosis -- increased radiotracer uptake in a WBC scan**

- Uninfected postsurgical wound
- Normal bone marrow activity
- Sterile hematomas
- Bleeding wounds
- Tumors

**Diagnosis:** Infected arteriovenous graft at the medial left arm

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# Arteriovenous (AV) graft infections

## Pathophysiology

Patients on chronic hemodialysis require vascular access, which can be achieved with central venous cannulation or via the creation of an autologous arteriovenous fistula or a fistula with an arteriovenous vascular prosthesis. One of the most serious complications of these grafts is a bacterial infection, which is usually caused by gram-positive bacteria, especially *Staphylococcus aureus*.

The majority of infections are thought to be caused by routine dialysis. The repetitive compromise of skin integrity allows an infection to reach the graft.

## Epidemiology

The frequency of vascular graft infections depends on the location of the graft, but it is

The frequency of vascular graft infections depends on the location of the graft, but it is reported to be between 1% and 6%.

## Clinical presentation

Most patients with AV graft infections present with low-grade fever, leukocytosis, a superficial surgical wound infection, and/or a draining sinus or nonhealing wound.

## Imaging features

- CT/ultrasound: These modalities can show a fluid collection around an infected graft, a thickened graft, graft occlusion, or an anastomotic aneurysm. CT can show air around an infected graft, which is highly specific for infection.
- F-18 FDG-PET/CT: Infection can appear as focal and/or inhomogeneous uptake greater than inactive muscle and fat or liver activity



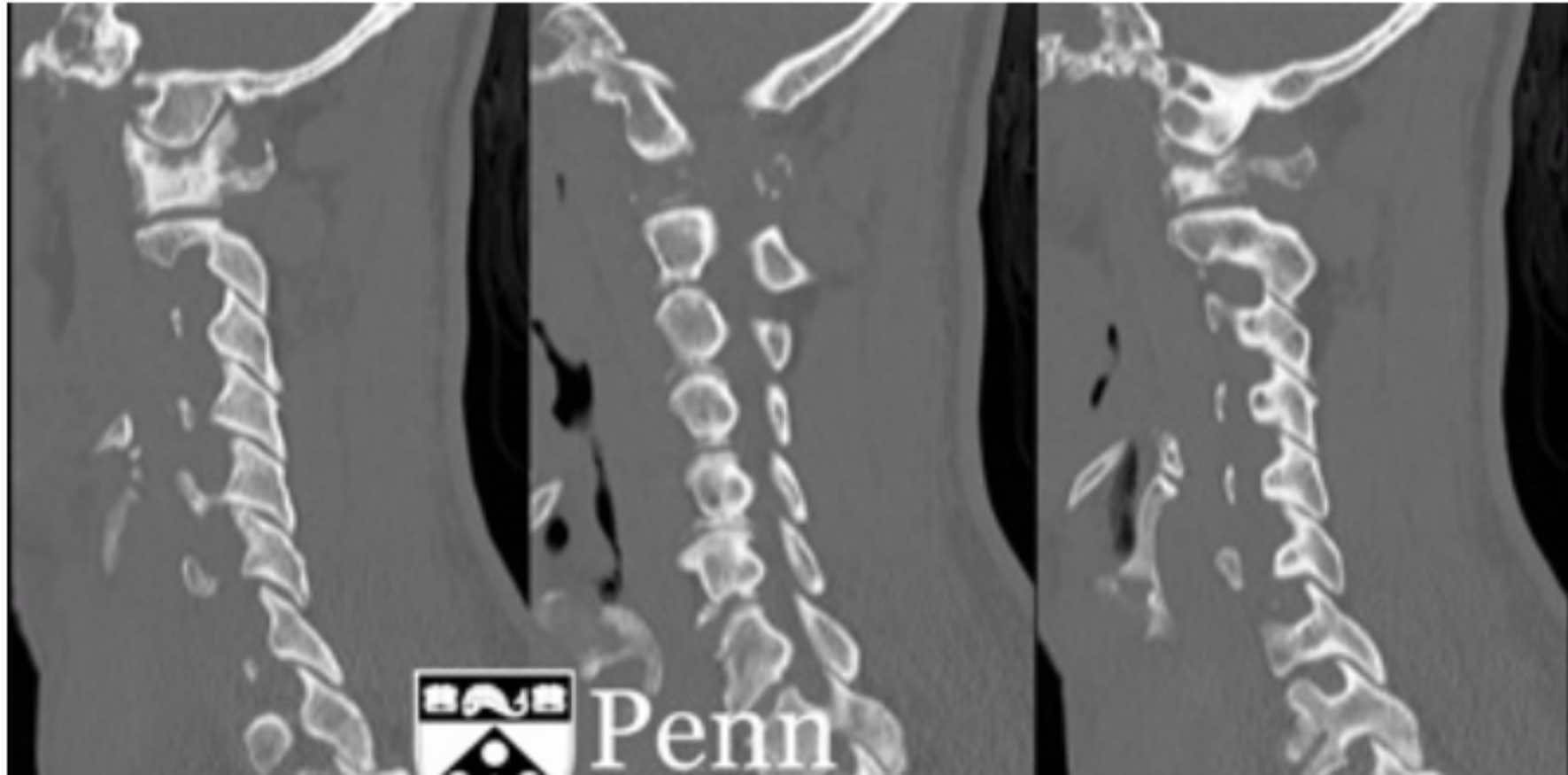
- F-18 FDG-PET/CT: Infection can appear as focal and/or inhomogeneous uptake greater than inactive muscle and fat or liver activity.
- Labeled white blood cell scan: An AV graft infection can appear as moderately increased uptake along the inner surface of the graft. An abscess or phlegmon usually presents with focal, asymmetrical uptake along or near the graft. Technetium-99m sulfur colloid can be useful to distinguish physiologic bone marrow uptake in indium-111 scans from osteomyelitis.

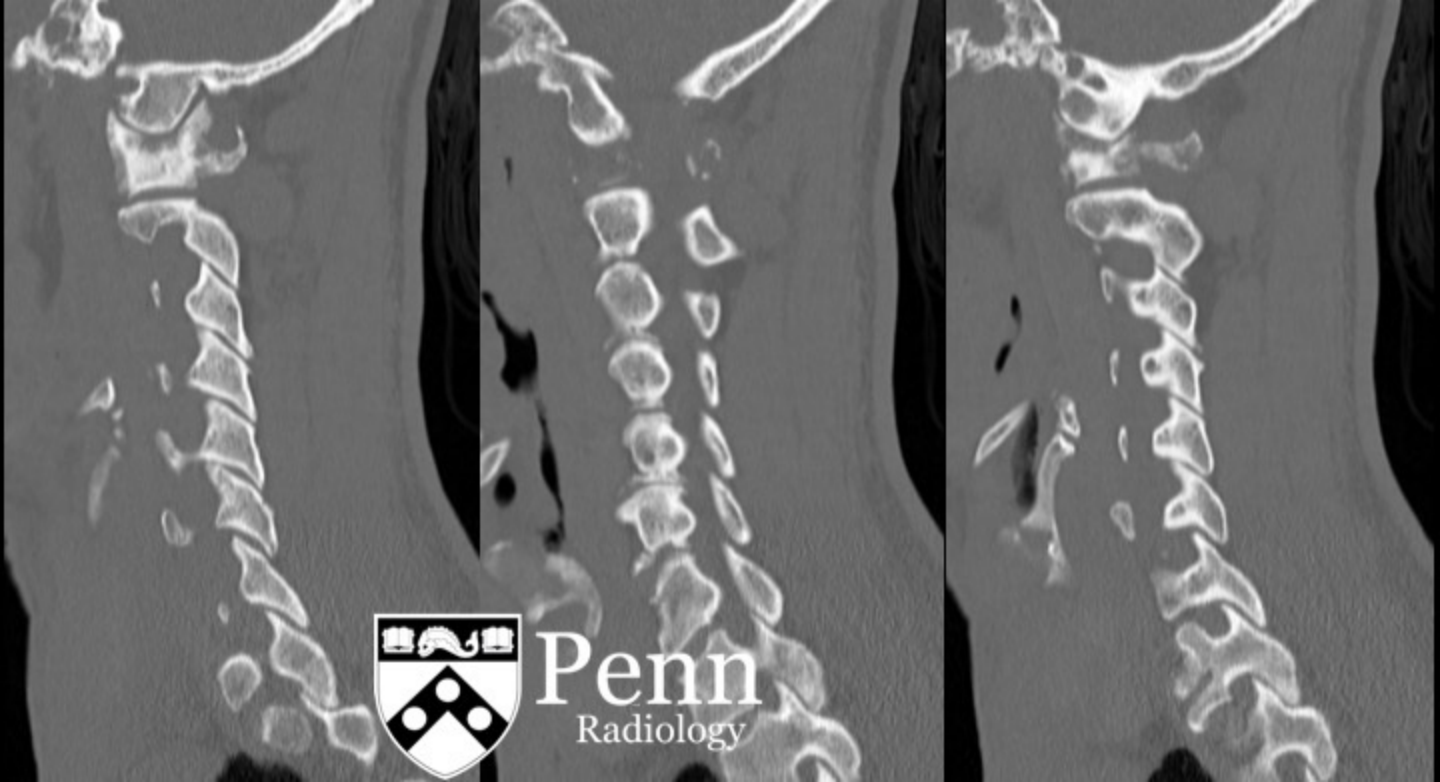
## Treatment

Many institutions treat dialysis prosthetic graft infections with total graft removal, including complete debridement of devitalized and infected tissues around the prosthesis, total graft excision, and in situ reconstruction with a new prosthesis or autologous vein or arterial allograft/homograft. Antibiotic treatment of AV graft infections also has had a major impact on morbidity of dialysis patients.

**History:** A 66-year-old man presents with neck pain and stiffness. He has a history of prostate cancer.

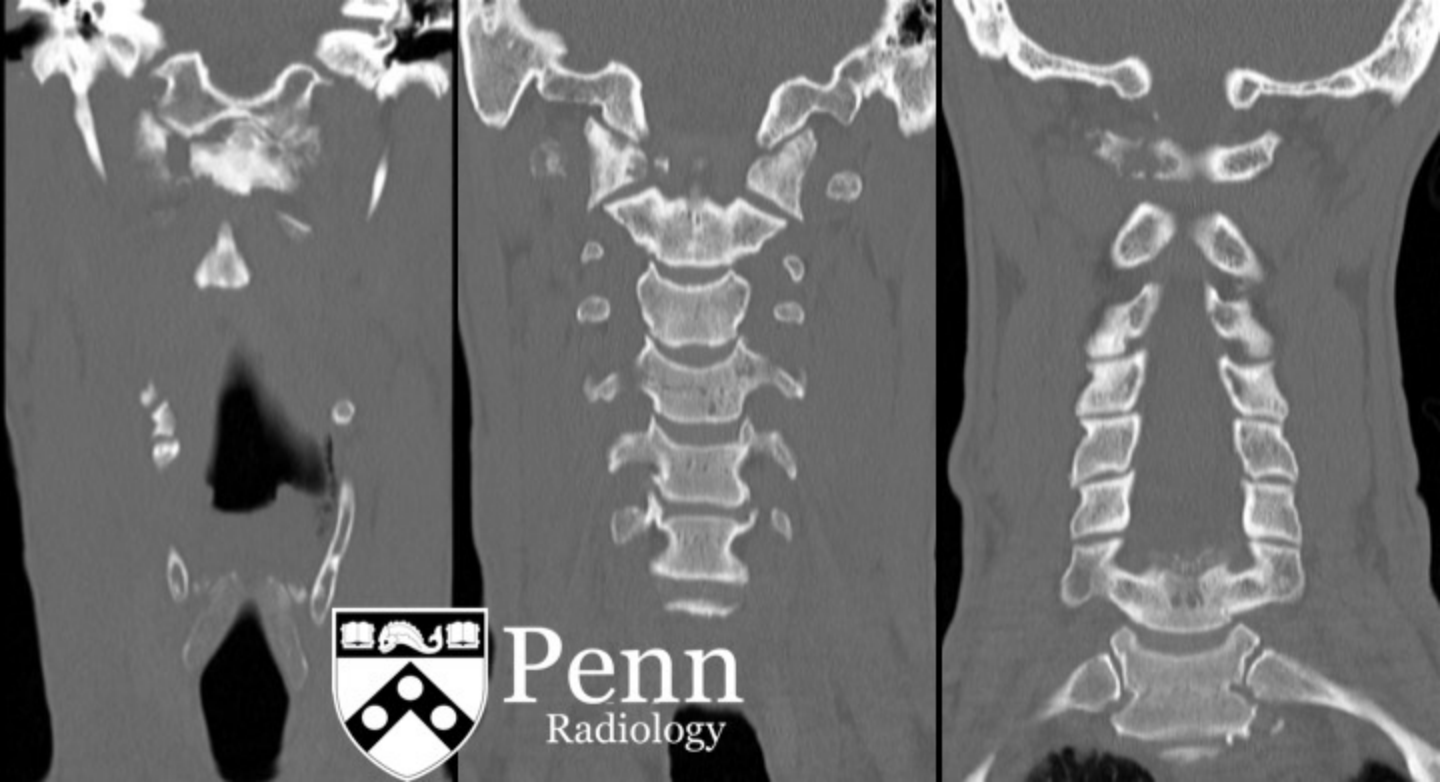
A CT scan of the cervical spine was performed. Axial, coronal, and sagittal images of the cervical spine in bone windows are shown below. Click to enlarge.



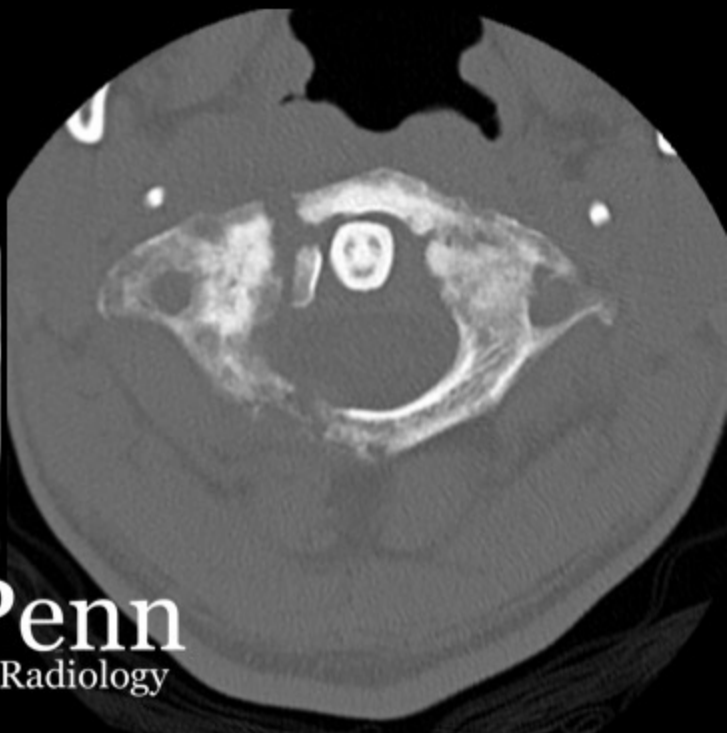


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## Which bone is fractured?

☐ Occipital condyle

☐ C1 (atlas)

☐ C2 (axis)

☐ C3

The question above accounts for 10% of your total score for this case

## Which bone is fractured?

☐ Occipital condyle

☒ C1 (atlas) (correct!)

☐ C2 (axis)

☐ C3

The question above accounts for 10% of your total score for this case

The question above accounts for 10% of your total score for this case.

**This is a pathologic fracture.**

☐ True

☐ False

The question above accounts for 10% of your total score for this case.

**What is the eponym for this type of fracture?**

☐ Hangman's fracture



The question above accounts for 10% of your total score for this case.

**This is a pathologic fracture.**

☒ True (correct!)

☐ False

The question above accounts for 10% of your total score for this case.

**What is the eponym for this type of fracture?**

☐ Hangman's fracture

The question above accounts for 10% of your total score for this case.

**What is the eponym for this type of fracture?**

☐ Hangman's fracture

☐ Jefferson fracture

☐ Clay-Shoveler's fracture

☐ Flexion teardrop fracture

The question above accounts for 10% of your total score for this case.

**What is the eponym for this type of fracture?**

☐ Hangman's fracture

☒ Jefferson fracture (correct!)

☐ Clay-Shoveler's fracture

☐ Flexion teardrop fracture

The question above accounts for 10% of your total score for this case.

**CT angiography (CTA) of the head and neck is indicated.**

☐ True

☐ False

The question above accounts for 10% of your total score for this case.

**Neurosurgery consultation is indicated.**

The question above accounts for 10% of your total score for this case.

**CT angiography (CTA) of the head and neck is indicated.**

☐ True

☒ False (correct!)

**[Explain this Answer]**

The question above accounts for 10% of your total score for this case.

**Neurosurgery consultation is indicated.**





X

☐ Flexion teardrop

The fracture does not traverse the foramen transversarium.

The question above

**CT angiograph**

☐ True

☒ False (correct)

[Explain this Answer]

The question above

**[Explain this Answer]**

The question above accounts for 10% of your total score for this case.

**Neurosurgery consultation is indicated.**

☐ True

☐ False

The question above accounts for 10% of your total score for this case.

**Which of the following is an appropriate follow-up imaging study?**

☐ Magnetic resonance imaging

**[Explain this Answer]**

The question above accounts for 10% of your total score for this case.

**Neurosurgery consultation is indicated.**

☒ True (correct!)

☐ False

The question above accounts for 10% of your total score for this case.

**Which of the following is an appropriate follow-up imaging study?**

☐ Magnetic resonance imaging

The question above accounts for 10% of your total score for this case.

**Which of the following is an appropriate follow-up imaging study?**

☐ Nuclear medicine bone scan

☐ Brain MRI

☐ MRI of the cervical, thoracic, and lumbar spine

☐ All of the above

The question above accounts for 10% of your total score for this case.

**Which of the following is an appropriate follow-up imaging study?**

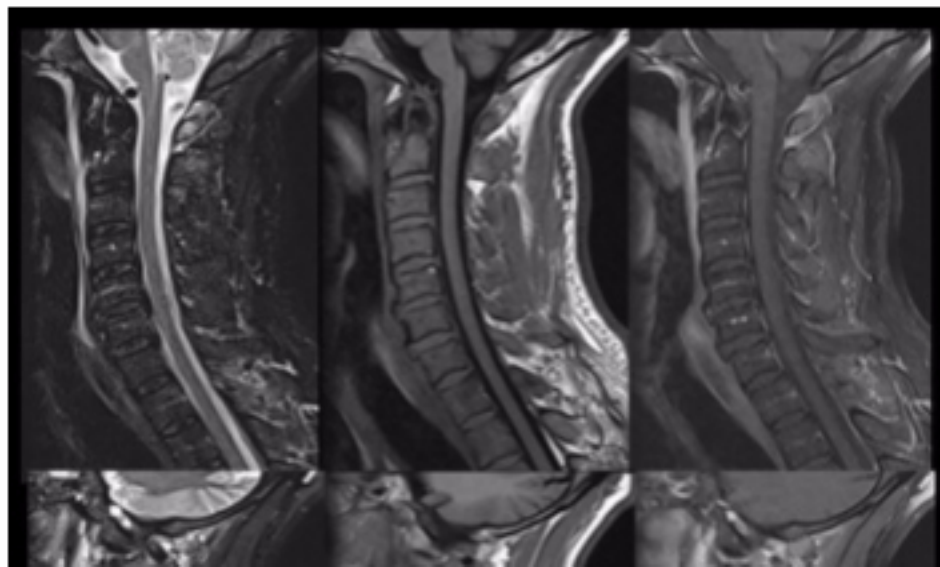
- ☐ Nuclear medicine bone scan
- ☐ Brain MRI
- ☐ MRI of the cervical, thoracic, and lumbar spine
- ☒ All of the above (correct!)

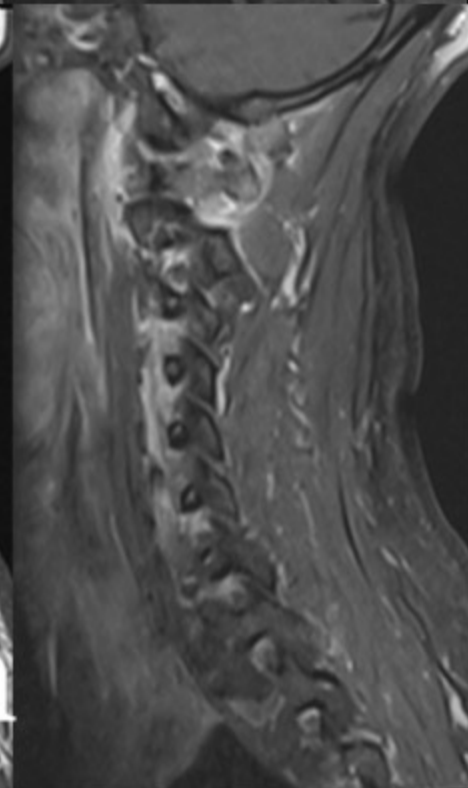
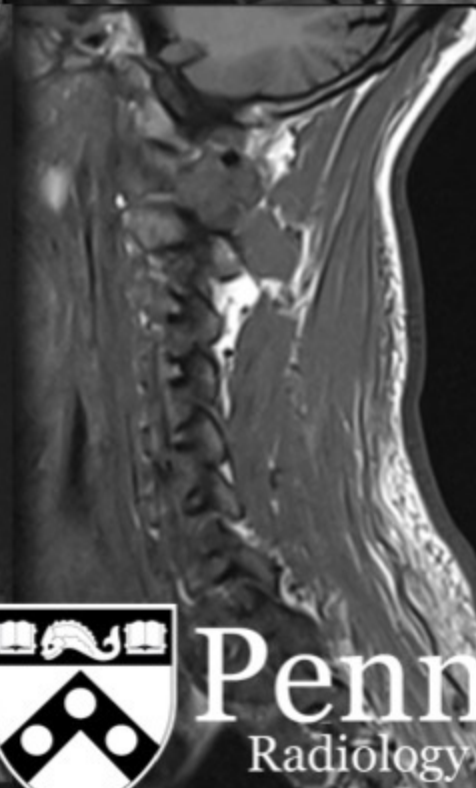
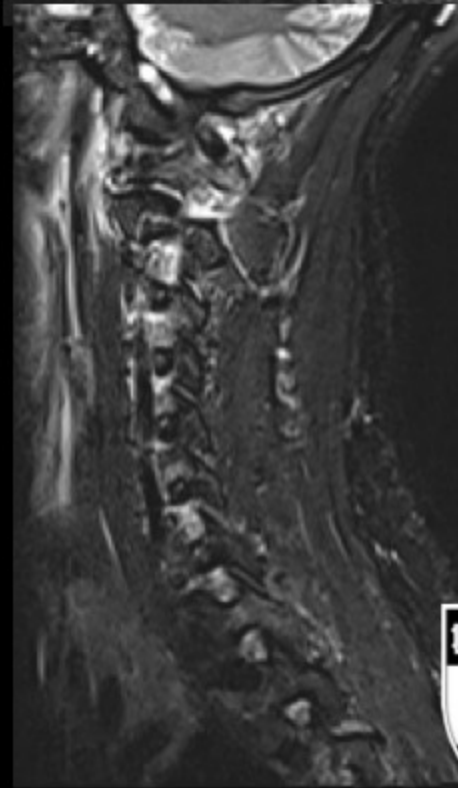
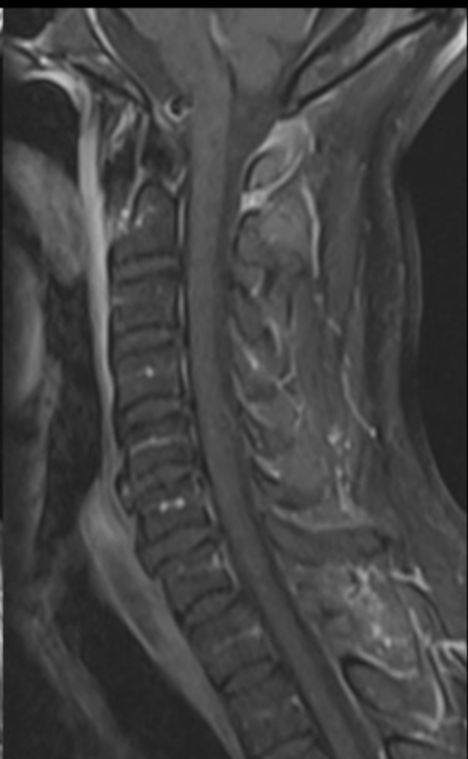
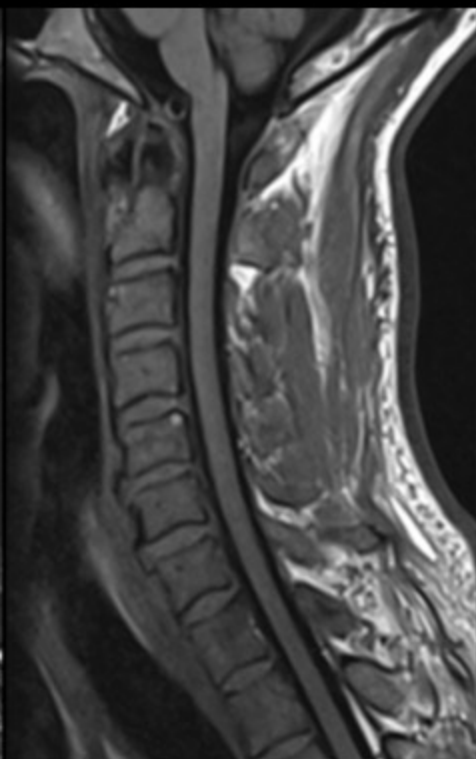


## Additional images

A CTA scan of the head and neck was not order given the fact that the foramen transversarium was not involved and the fracture was not traumatic.

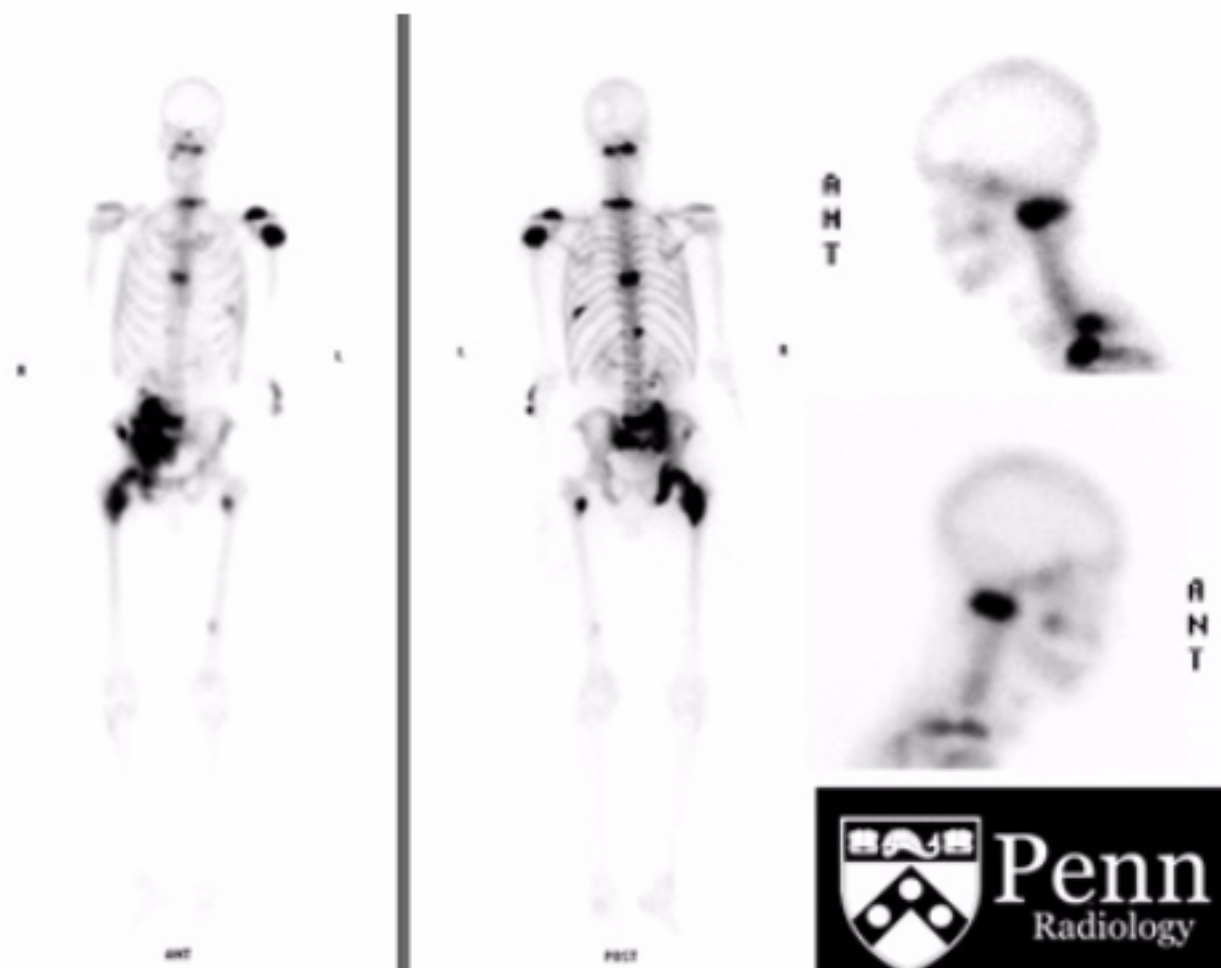
An MRI scan of the brain and entire spine was obtained. Sagittal short tau inversion-recovery (STIR), T1-weighted fluid-attenuated inversion-recovery (FLAIR), and T1-weighted postcontrast images of the cervical spine are shown below. Click to enlarge.

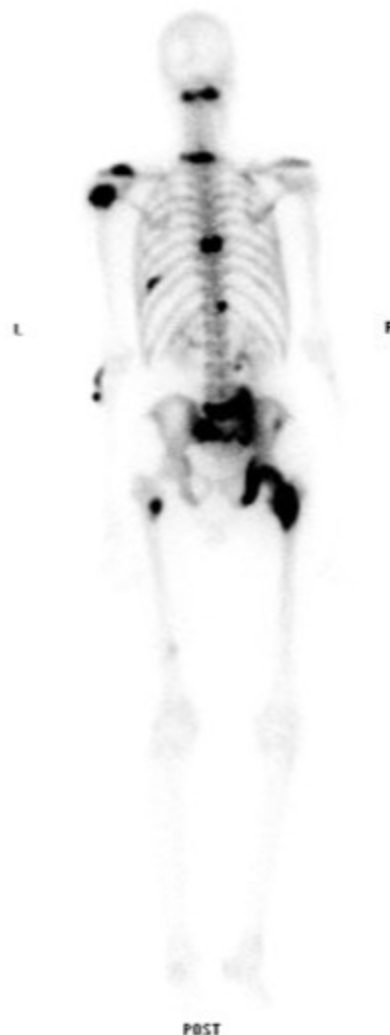




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A nuclear medicine bone scan also was performed. Whole-body anterior and posterior images and also spot images of the skull approximately three hours after the administration of 27.4 mCi of technetium-99m methylene diphosphonate (Tc-99m MDP) are shown below. Click to enlarge.





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**There is abnormal signal in the spinal cord.**

☐ True

☐ False

The question above accounts for 10% of your total score for this case.

**There is retropharyngeal fluid and mild enhancement.**



**There is abnormal signal in the spinal cord.**

☐ True

☒ False (correct!)

The question above accounts for 10% of your total score for this case.

**There is retropharyngeal fluid and mild enhancement.**

The question above accounts for 10% of your total score for this case.

**There is retropharyngeal fluid and mild enhancement.**

☐ True

☐ False

The question above accounts for 10% of your total score for this case.

**There are widespread osseous metastases.**

☐ True

The question above accounts for 10% of your total score for this case.

**There is retropharyngeal fluid and mild enhancement.**

☒ True (the correct answer)

☐ False (wrong) <- your answer

The question above accounts for 10% of your total score for this case.

**There are widespread osseous metastases.**

☐ True

The question above accounts for 10% of your total score for this case.

**There are widespread osseous metastases.**

☐ True

☐ False

The question above accounts for 10% of your total score for this case.

**What is the most appropriate therapy for this patient?**

☐ Chemotherapy and hard collar immobilization

The question above accounts for 10% of your total score for this case.

**There are widespread osseous metastases.**

☒ True (correct!)

☐ False

The question above accounts for 10% of your total score for this case.

**What is the most appropriate therapy for this patient?**

☐ Chemotherapy and hard collar immobilization



The question above accounts for 10% of your total score for this case.

**What is the most appropriate therapy for this patient?**

- ☐ Chemotherapy and hard collar immobilization
- ☐ Radiation therapy and hard collar immobilization
- ☐ Spinal fusion surgery

The question above accounts for 10% of your total score for this case.

The question above accounts for 10% of your total score for this case.

**What is the most appropriate therapy for this patient?**

☐ Chemotherapy and hard collar immobilization (wrong) <- your answer

☒ Radiation therapy and hard collar immobilization (the correct answer)

☐ Spinal fusion surgery

The question above accounts for 10% of your total score for this case.

cervical spine with a minimally displaced pathologic fracture of the right anterior and posterior arches of C1 is seen. There is associated marrow heterogeneity and enhancement, likely representing metastasis. No definite evidence of epidural extension of neoplasm is seen. Vertebral alignment is otherwise anatomic. There is a small to moderate amount of prevertebral edema. Associated ill-defined enhancement in the prevertebral soft tissues is likely reactive. The cervical spinal cord is without definite signal abnormality.

- **Nuclear medicine bone scan:** There are widespread osseous foci of abnormal radiotracer uptake consistent with osseous metastases. This includes C1 and T1 vertebral bodies, left posterior eighth rib, anterior left fifth rib, lower thoracic spine, left scapula and proximal humerus, multiple right pelvic bones, right proximal femur, and proximal and distal left femur.

## Differential diagnosis

- Pathologic Jefferson (C1) fracture
- Traumatic Jefferson (C1) fracture



in the prevertebral soft tissues is likely reactive. The cervical spinal cord is without definite signal abnormality.

- **Nuclear medicine bone scan:** There are widespread osseous foci of abnormal radiotracer uptake consistent with osseous metastases. This includes C1 and T1 vertebral bodies, left posterior eighth rib, anterior left fifth rib, lower thoracic spine, left scapula and proximal humerus, multiple right pelvic bones, right proximal femur, and proximal and distal left femur.

## Differential diagnosis

- Pathologic Jefferson (C1) fracture
- Traumatic Jefferson (C1) fracture
- Pathologic Hangman's (C2) fracture
- Odontoid/dens fracture
- Clay-Shoveler's fracture
- Osteonecrosis
- Multiple myeloma

proximal and distal left femur.

## Differential diagnosis

- Pathologic Jefferson (C1) fracture
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- Pathologic Hangman's (C2) fracture
- Odontoid/dens fracture
- Clay-Shoveler's fracture
- Osteonecrosis
- Multiple myeloma
- Primary bone tumor

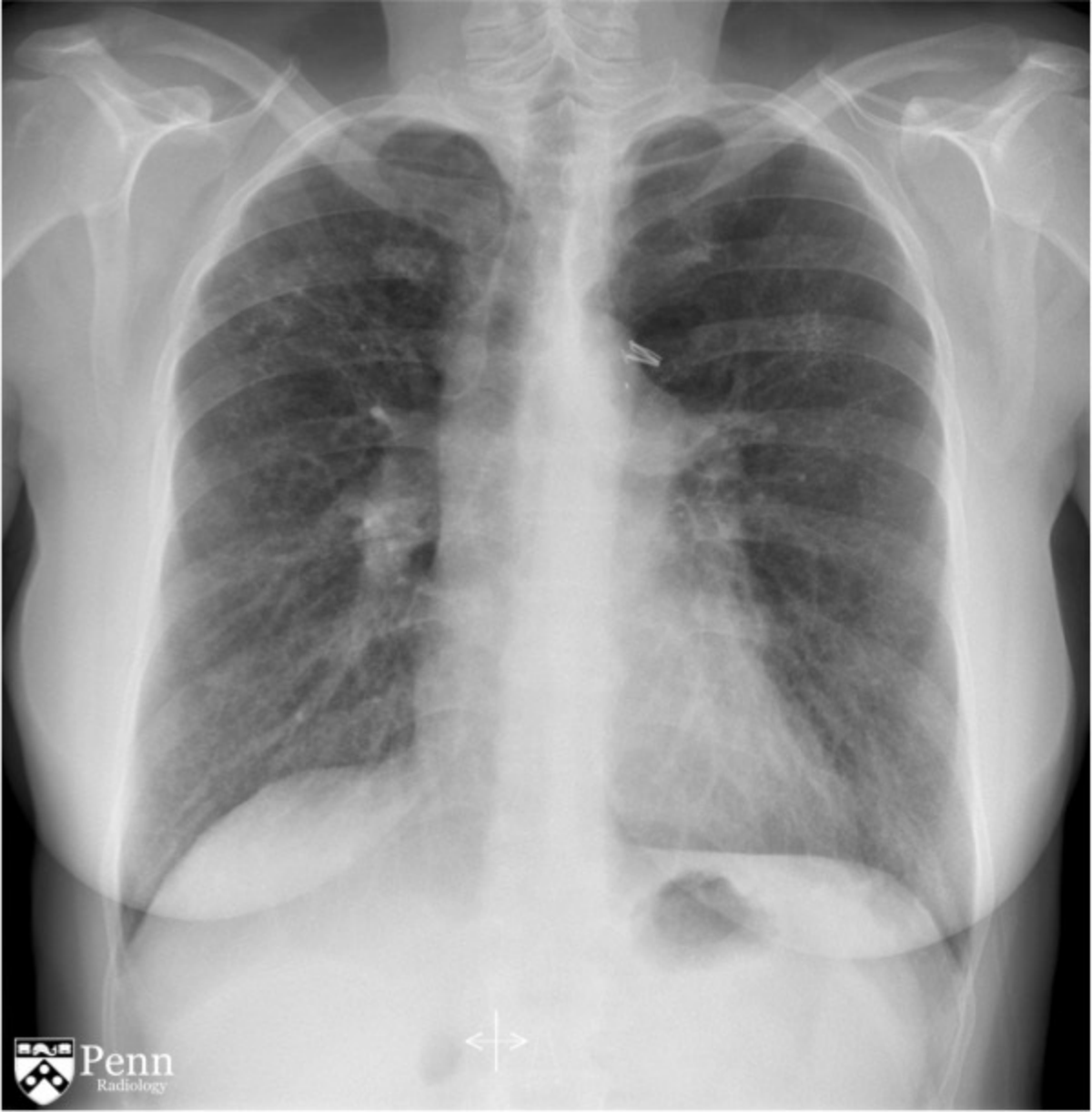
**Diagnosis:** Pathologic Jefferson (C1) fracture

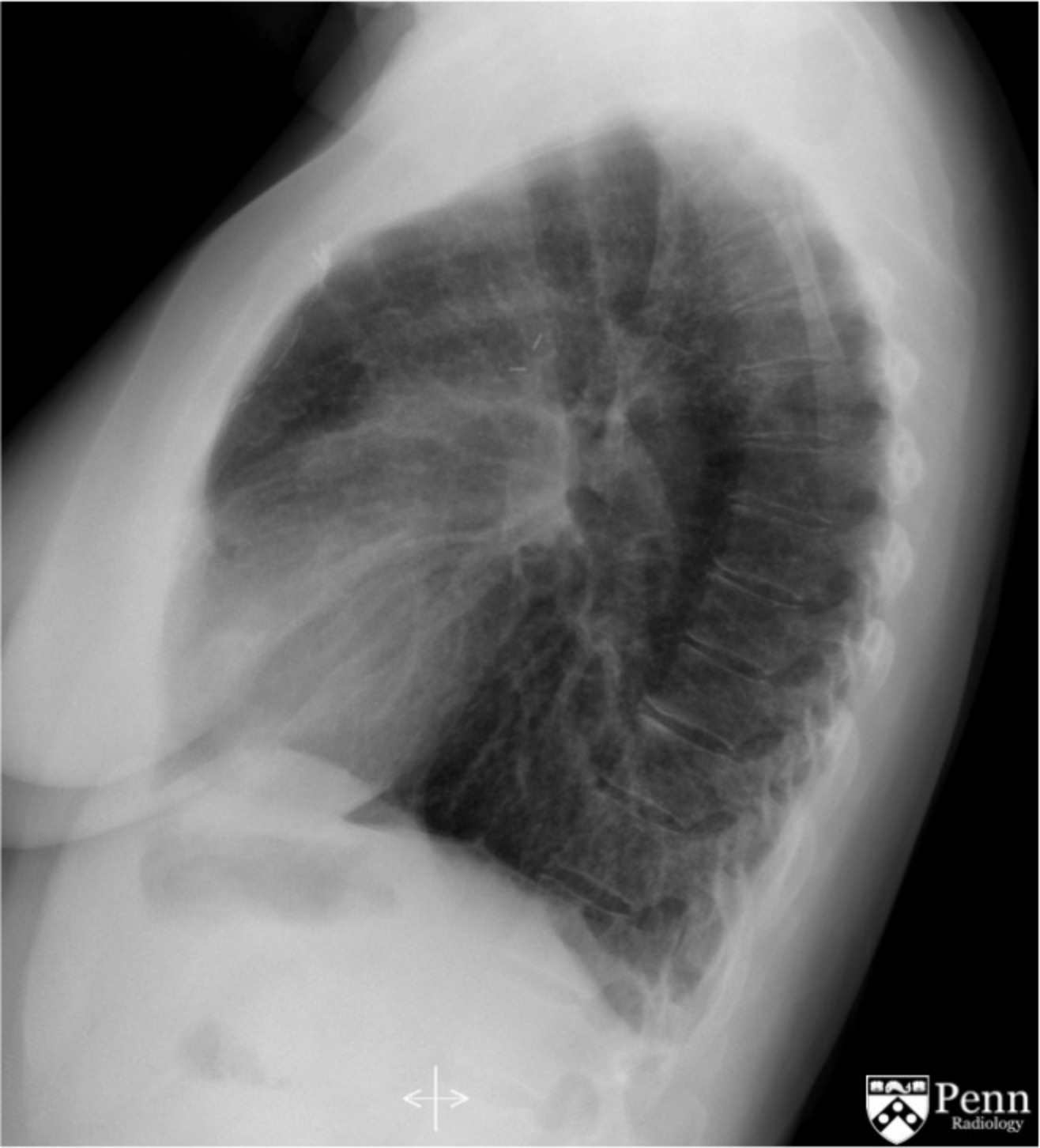


**History:** A 52-year-old woman undergoing a workup for lung transplantation. She has a long-term history of progressive, severe shortness of breath and dyspnea.

Posteroanterior and lateral chest radiographs are shown below. Click to enlarge.







**The pulmonary arteries are enlarged.**

☐ True

☐ False

The question above accounts for 15% of your total score for this case.

**There is relative increased lucency of the upper and mid lung zones.**

☐ True

☐ False

**The pulmonary arteries are enlarged.**

☒ True (correct!)

☐ False

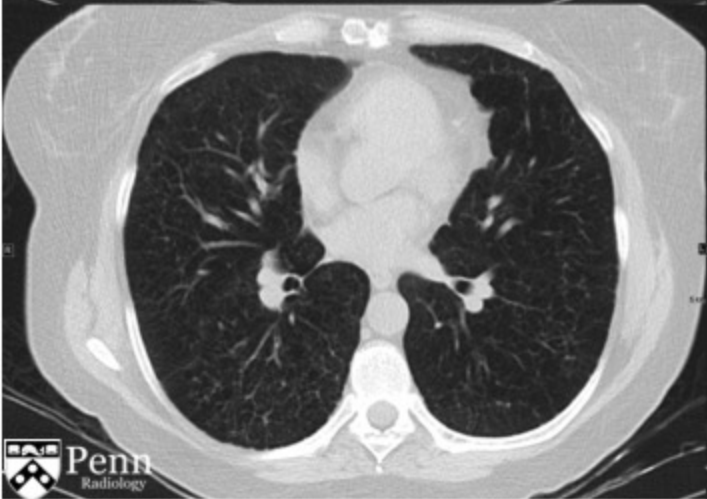
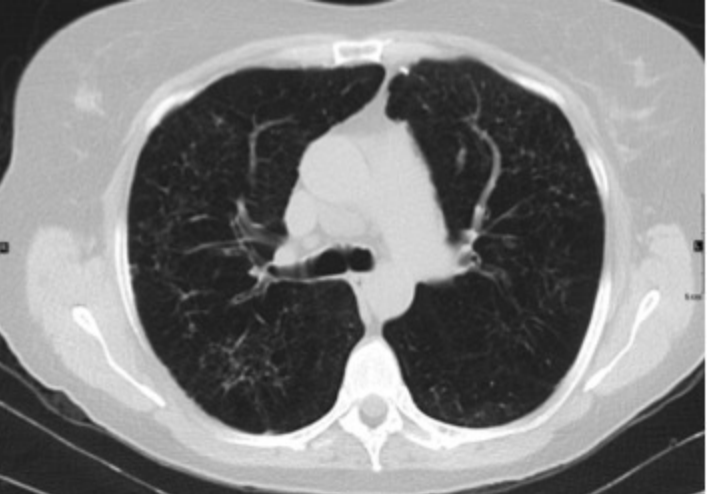
**[Explain this Answer]**

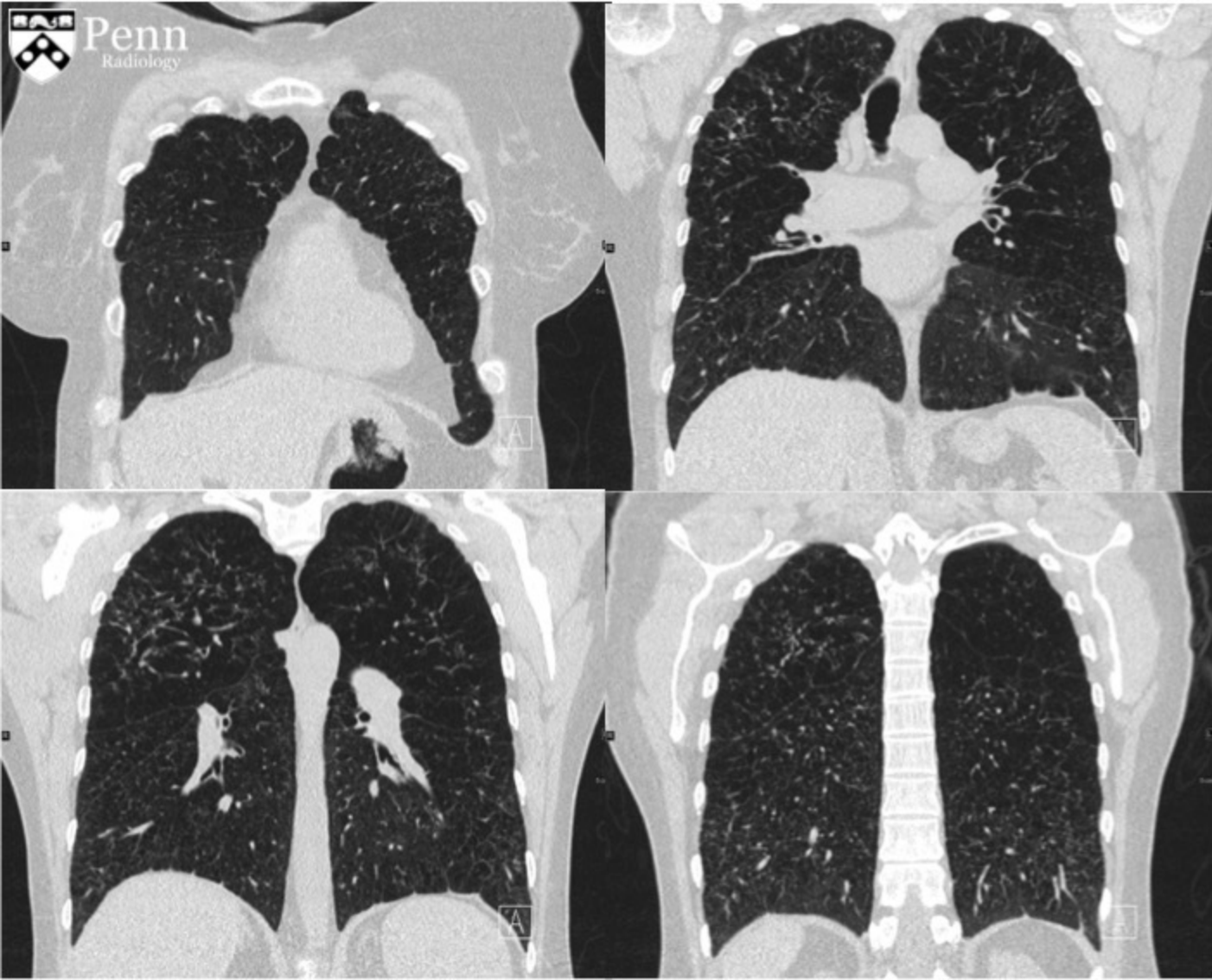
The question above accounts for 15% of your total score for this case.

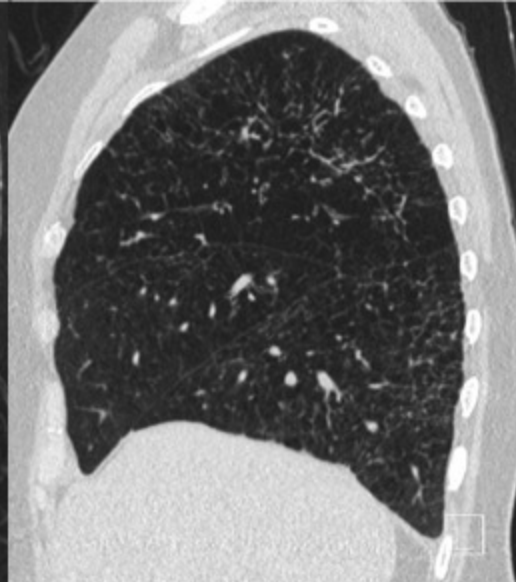
**There is relative increased lucency of the upper and mid lung zones.**

☒ True (correct!)











**What is the major finding?**

☐ Infection

☐ Malignancy

☐ Cystic lung disease

☐ Pneumothorax



**What is the major finding?**

☐ Infection

☐ Malignancy

☒ Cystic lung disease (correct!)

☐ Pneumothorax



### [Explain this Answer]

The question above accounts for 14% of your total score for this case.

**There is relative sparing of the lung bases.**

☐ True

☐ False

The question above accounts for 14% of your total score for this case.

[VIEW YOUR SCORE](#)

The question above accounts for 14% of your total score for this case.

**There is relative sparing of the lung bases.**

☒ True (correct!)

☐ False

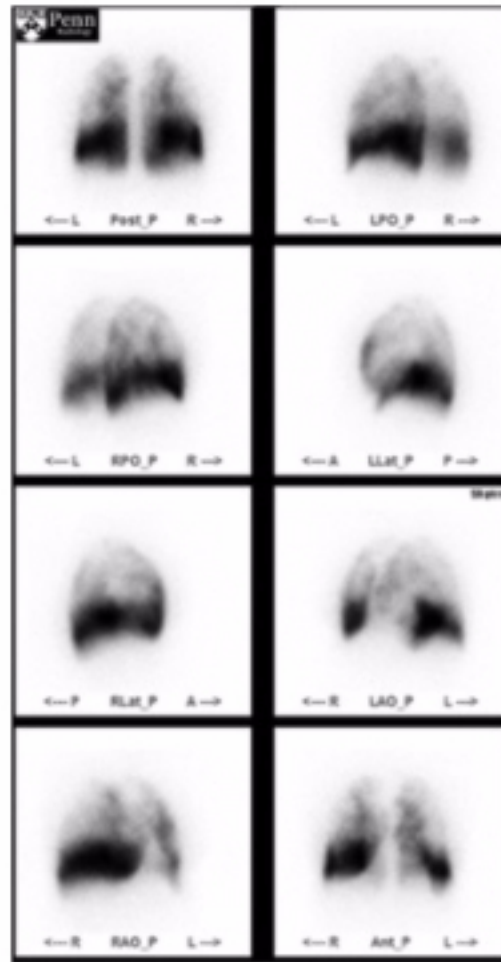
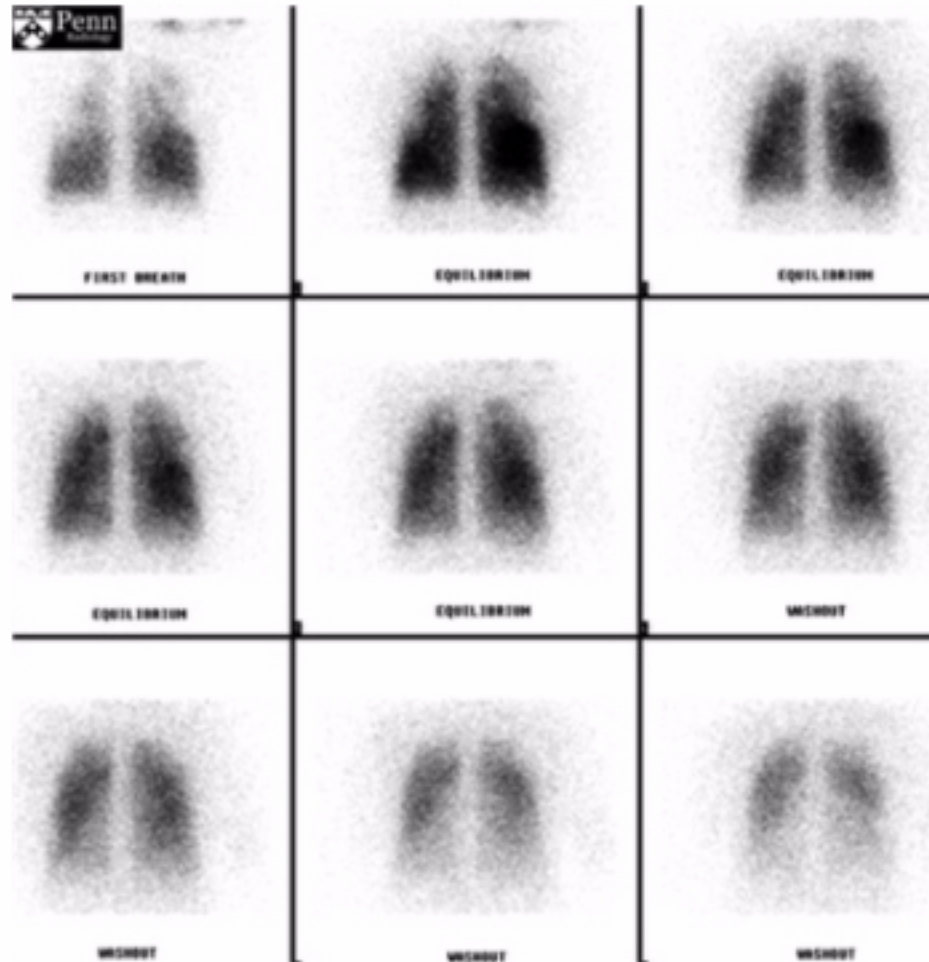
**[Explain this Answer]**

The question above accounts for 14% of your total score for this case.

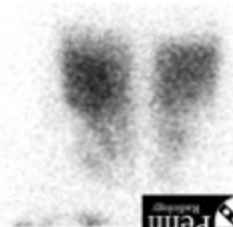
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# V/Q scan images

The patient underwent a ventilation/perfusion (V/Q) scan. Shown below are xenon-133 (Xe-133) ventilation images in the posterior projection during single-breath, equilibrium, and washout phases and technetium-99m microaggregated albumin (Tc-99m MAA) perfusion scan images. Click to enlarge.



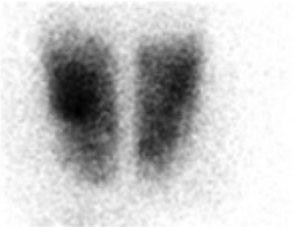
FIRST BREATH



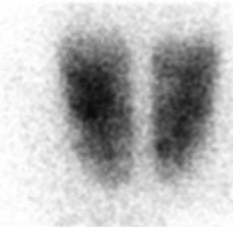
EQUILIBRIUM



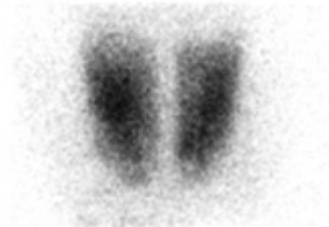
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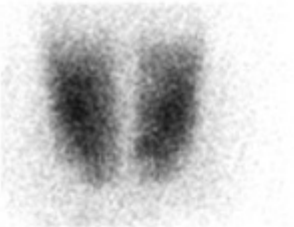
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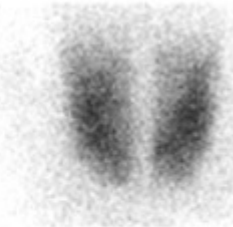
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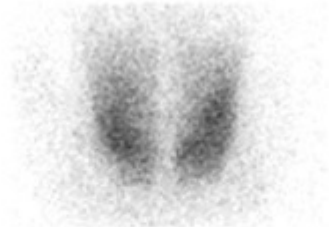
WASHOUT



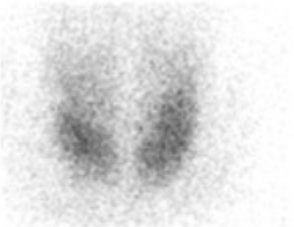
WASHOUT



WASHOUT



WASHOUT





<--- L Post\_P R --->



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50 pax



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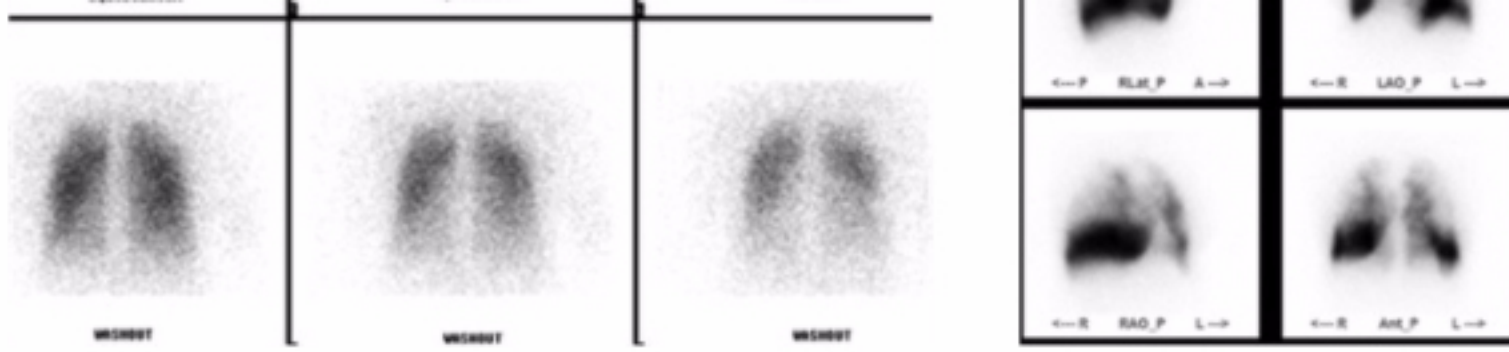


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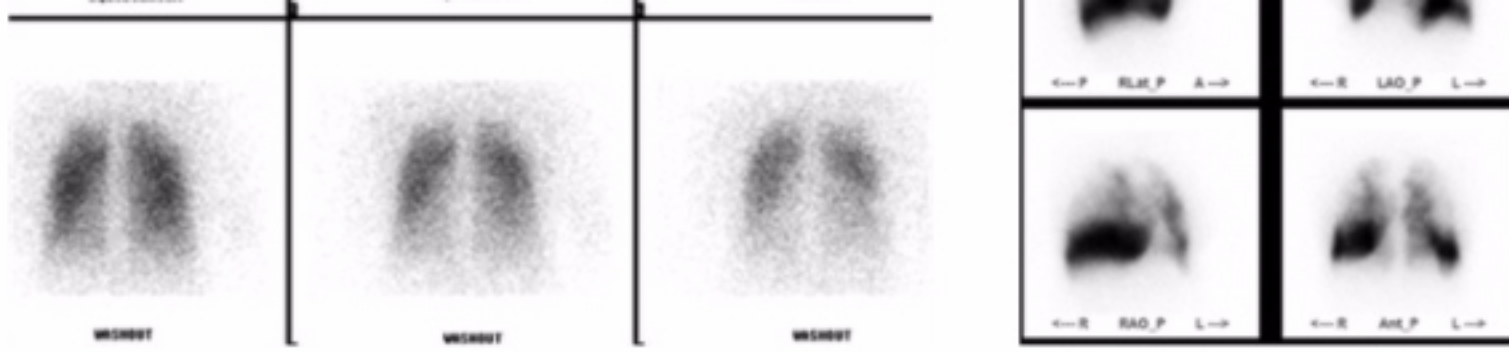
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**Which of the following is seen on the Xe-133 ventilation images?**

- ☐ Air trapping in the lung apices during the washout phase
- ☐ Uniform distribution of radiotracer throughout the study
- ☐ Air trapping in the lung bases during the washout phase



**Which of the following is seen on the Xe-133 ventilation images?**

- ☒ Air trapping in the lung apices during the washout phase (correct!)
- ☐ Uniform distribution of radiotracer throughout the study
- ☐ Air trapping in the lung bases during the washout phase



X

Radiotracer is noted in the lung apices at the end of the washout phase due to air trapping.

Which of the fo

☒ Air trapping in

☐ Uniform distri

☐ Air trapping in

[[Explain this answer](#)]

The question above accounts for 14% of your total score for this case.

**The underlying lung disease is most commonly associated with which of the following?**

☐ Chronic opioid abuse

☐ Smoking history

☐ Tuberculosis infection

☐ Work-related chemical exposure

[[Explain this answer](#)]

The question above accounts for 14% of your total score for this case.

**The underlying lung disease is most commonly associated with which of the following?**

☐ Chronic opioid abuse

☒ Smoking history (correct!)

☐ Tuberculosis infection

☐ Work-related chemical exposure



[Explain this / Ans]

The question above



**The underlying  
following?**

☐ Chronic opioi

☒ Smoking histo

☐ Tuberculosis

☐ Work-related

The only consistent epidemiological association of this disease entity is with cigarette smoking.

h which of the

### [Explain this Answer]

The question above accounts for 14% of your total score for this case.

**Which of the following is seen on the perfusion study?**

- ☐ Large wedge-shaped perfusion defects at the lung bases
- ☐ Uniform perfusion
- ☐ Artificially decreased perfusion at the lung apices
- ☐ Large areas of decreased radiotracer uptake within the upper and mid lung zones bilaterally

### [Explain this Answer]

The question above accounts for 14% of your total score for this case.

**Which of the following is seen on the perfusion study?**

- ☐ Large wedge-shaped perfusion defects at the lung bases
- ☐ Uniform perfusion
- ☐ Artifactually decreased perfusion at the lung apices
- ☒ Large areas of decreased radiotracer uptake within the upper and mid lung zones bilaterally (correct!)

Which of the following is seen on the perfusion study?

☐ Large wedge-

☐ Uniform perfu

☐ Artifactually d

☒ Large areas of  
bilaterally (corre



X

There are large areas of decreased radiotracer uptake within the upper and mid lung zones bilaterally corresponding to regions of severe cystic lung disease on CT. This is not due to artifact.

lung zones

[Explain this Answer]

The question above



## Findings

- **Radiographs:**

- There is relatively increased lucency of the upper and mid lung zones bilaterally due to underlying cystic lung disease.
- Enlarged pulmonary arteries are seen.
- There are also surgical changes from a previous lung biopsy.

- **CT:** There is severe diffuse cystic lung disease predominantly involving the upper and mid lung bilaterally with relative sparing of the bases.

- **V/Q perfusion scan:**

- Xe-133 ventilation scan: Initially demonstrates decreased radiotracer localization to bilateral upper lobes. Radiotracer eventually localizes within the upper lobes on equilibrium images and is retained in the upper lobes on washout images.
- Tc-99m MAA perfusion scan: Large areas of decreased radiotracer distribution in both upper and mid lung zones bilaterally. These perfusion defects are matched with the ventilation defects on the initial single-breath ventilation images.



with the ventilation defects on the initial single breath ventilation images.

## Differential diagnosis

- Pulmonary Langerhans cell histiocytosis
- Bullous emphysema
- Lymphangioleiomyomatosis
- Pneumocystis carinii pneumonia

**Diagnosis:** Pulmonary Langerhans cell histiocytosis

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[3](#)

[4](#)

[5](#)

[next »](#)

## Key points

### Pulmonary Langerhans cell histiocytosis (LCH)

#### Pathophysiology

- Pathogenesis of Langerhans cell histiocytosis remains poorly understood.
- Studies have demonstrated that LCH lesions represent clonal proliferation. Immunologic factors, viral infections, and somatic mutations have been proposed as potential causes of LC proliferation.
- It has also been suggested that LCH cells represent a form of activated Langerhans cells.
- Immunologic dysregulation also has been proposed as a cause of LCH due to the cytokine content within LCH lesions.

#### Epidemiology

## **Epidemiology**

- The precise incidence and prevalence are unknown.
- Lung biopsy studies of patients with interstitial lung disease identified LCH in only 5% of cases, suggesting that it is relatively rare. Likely underestimates true incidence.
- Only consistent epidemiologic association is with cigarette smoking, as more than 90% of patients have a history of smoking.
- Occurs mostly among Caucasians.
- Typically affects young adults (20-40 years).

## **Clinical presentation**

- Most patients are symptomatic, but 25% of patients are asymptomatic.
- Usual symptom duration is one to three months before diagnosis.
- Most common symptoms are listed below:
  - Nonproductive cough (50% to 70%)



- Dyspnea (33% to 87%)
- Less common symptoms include fatigue, weight loss, chest pain (frequently pleuritic), fever, wheezing, hemoptysis, and constitutional symptoms.
- Physical examination findings may include diminished breath sounds, rales, wheezing, and digital clubbing.
- Variable pulmonary function tests with obstructive, restrictive, and mixed patterns described.
- Most common pulmonary function abnormality is reduced carbon monoxide diffusing capacity.

## Imaging features

- Radiographs/CT:
  - Early disease: The most common findings are symmetric and bilateral small pulmonary nodules predominantly in the upper and middle lung zones with sparing of the lung bases.
  - Disease progression: More reticulonodular predominance

pulmonary nodules predominantly in the upper and middle lung zones with sparing of the lung bases.

- Disease progression: More reticulonodular predominance
- Further progression: Predominance of cystic changes that may mimic bullous emphysema or lymphangioleiomyomatosis
- End-stage disease: coarse reticular opacities usually in the upper and middle lung zones that may progress to honeycombing.
- May show increased lung volumes.
- CT:
  - CT is the most sensitive modality.
  - Early-phase disease: Shows predominantly nodular pattern of disease.
  - Late-phase disease: Shows predominantly thin-walled cysts.
  - Typical sequence of disease progression seen: nodules -> cavitory nodules -> thick-walled cysts -> thin-walled cysts -> confluent cysts

## Differential diagnoses



## Differential diagnoses

- Early-stage disease: Sarcoidosis, silicosis, metastases, military tuberculosis, hematogenous infections
- Late-stage disease: Bullous emphysema, lymphangioleiomyomatosis, pneumocystis carinii pneumonia

## Treatment

- Treatment includes smoking cessation.
- Glucocorticoid therapy is used for recent-onset symptomatic nodular pulmonary LCH.
- Cytotoxic agents (vinblastine, methotrexate, etc.) in combination with glucocorticoid therapy are used for severe multisystemic LCH.
- Lung transplantation has been performed on patients with severe respiratory failure or major pulmonary hypertension.
- Recurrence is associated with resumption of smoking and extrapulmonary involvement.

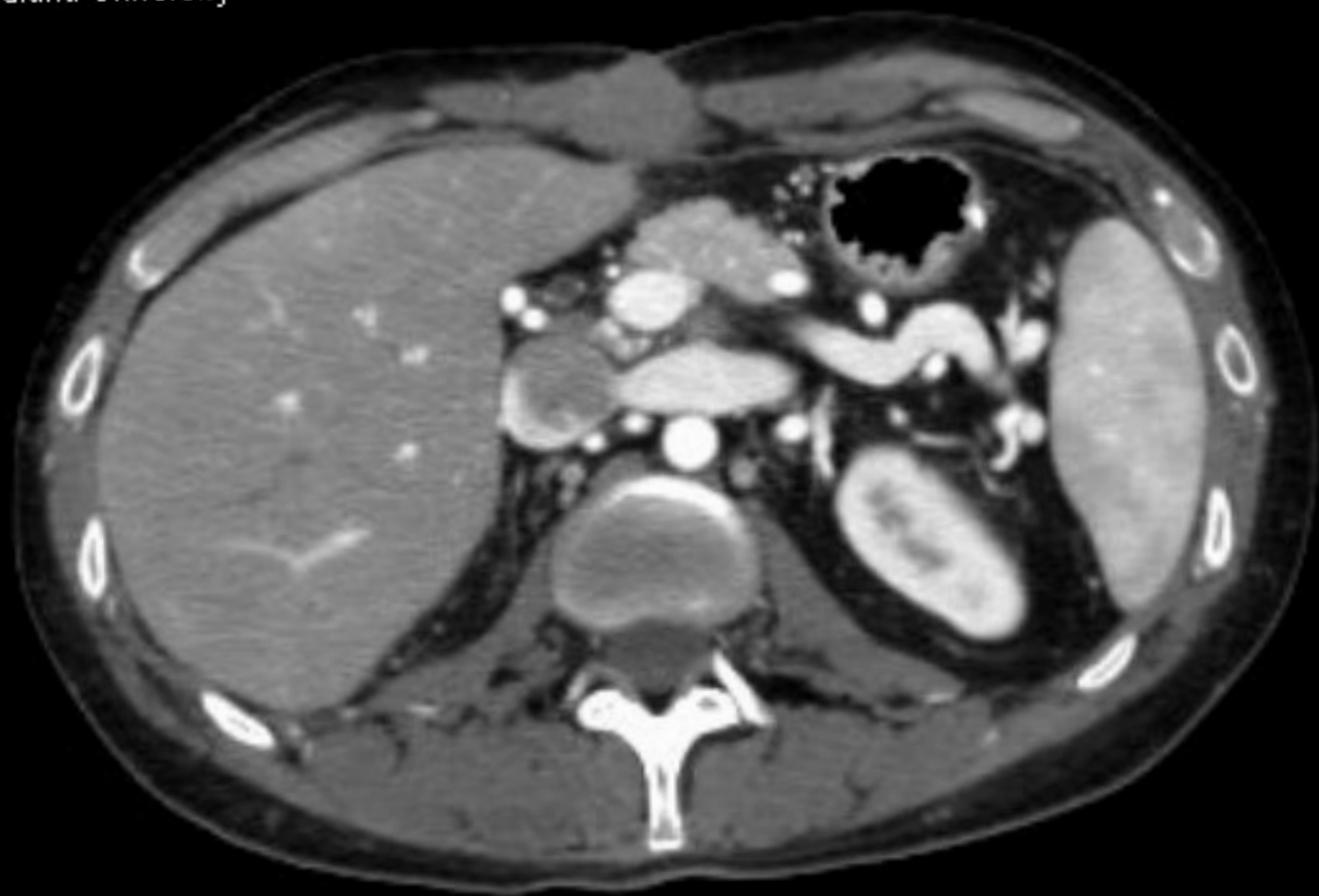
## References

**History:** Adult presents with abdominal pain.

CT images are shown below. Click to enlarge.











**Which choice best localizes the salient abnormality?**

☐ Pancreas

☐ Portal vein

☐ Spleen

☐ Inferior vena cava

☐ Abdominal wall

**Which choice best localizes the salient abnormality?**

☐ Pancreas

☐ Portal vein

☐ Spleen

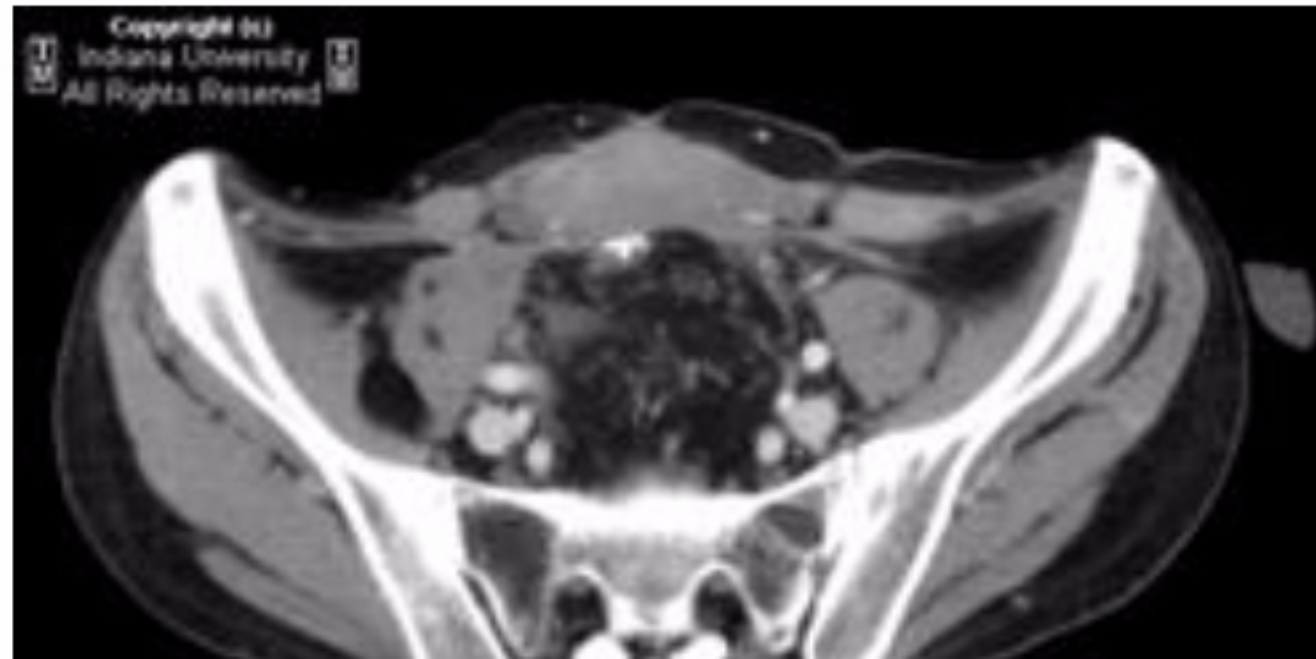
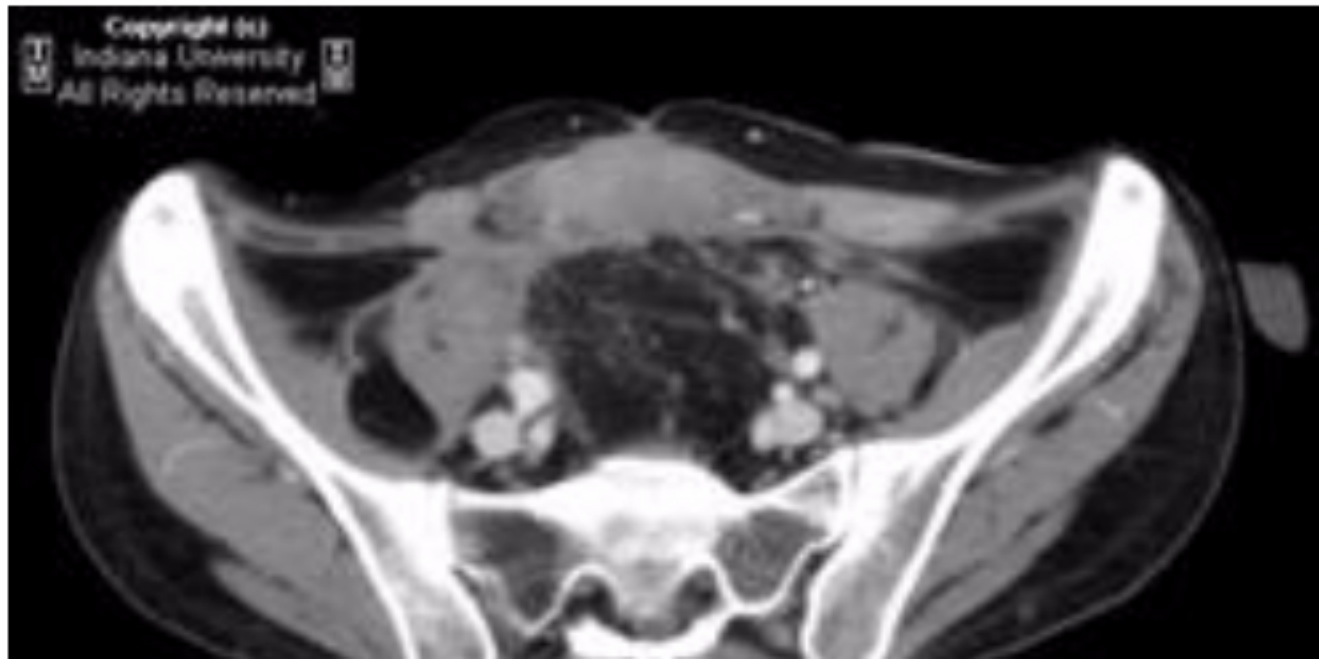
☐ Inferior vena cava

☒ Abdominal wall (correct!)

## Additional history and images

**Additional clinical history:** The patient is status post multiple small-bowel resections and subtotal colectomy.

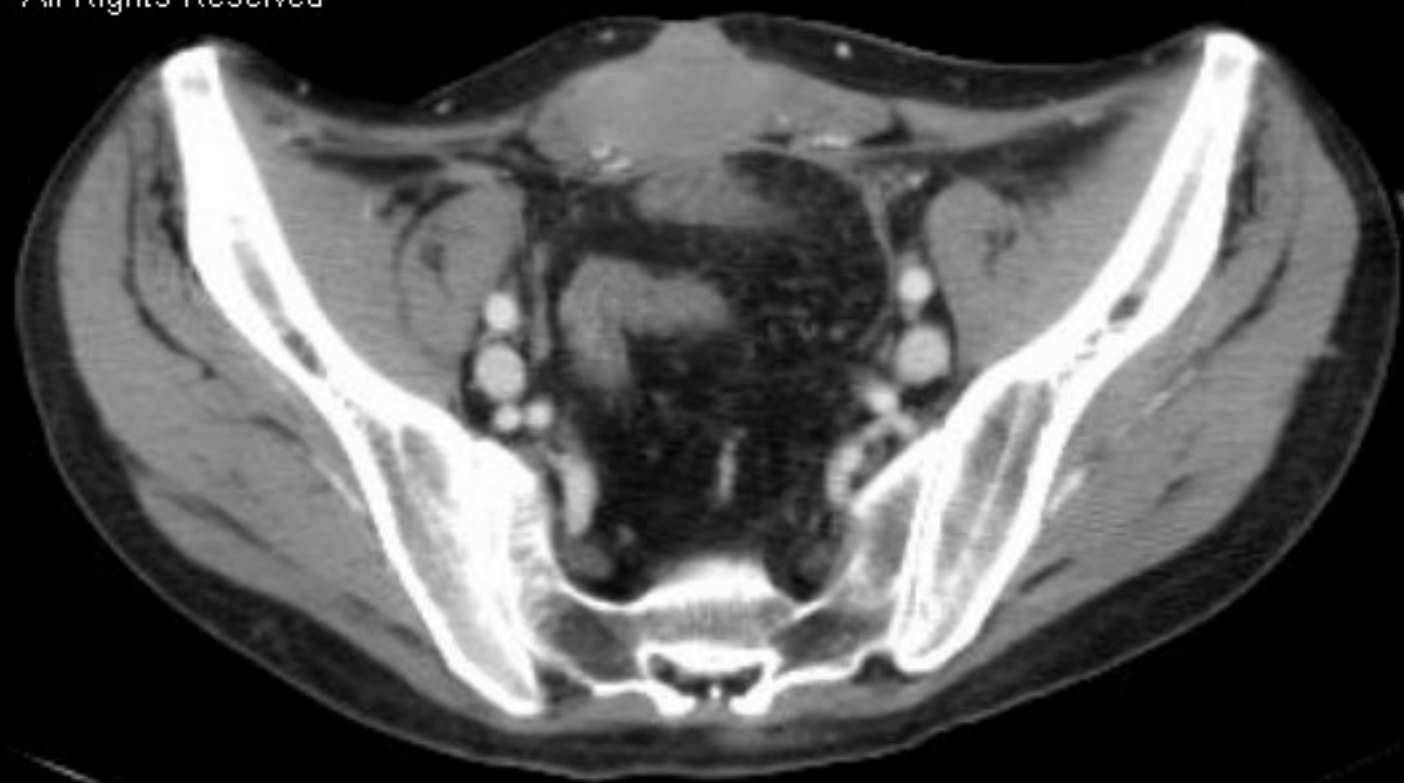
Below are images from the pelvis. Click to enlarge.













## What is the diagnosis?

☐ Melanoma metastases

☐ Desmoid tumors

☐ Rhabdomyosarcoma

☐ Necrotizing fasciitis

☐ Old hematomas

## What is the diagnosis?

☐ Melanoma metastases

☒ Desmoid tumors (correct!)

☐ Rhabdomyosarcoma

☐ Necrotizing fasciitis

☐ Old hematomas



X

What is the dia

Patient has had multiple bowel resections for desmoids.

☐ Melanoma me

☒ Desmoid tum

☐ Rhabdomyos

☐ Necrotizing fa

☐ Old hematoma



## Findings

There are multiple soft-tissue masses in the anterior abdominal muscles. There is some enhancement of the most dominant mass extending down the inferior rectus abdominus muscle.

## Differential diagnosis -- mesenteric masses

- Desmoid tumor
- Carcinoid tumor
- Lymphoma
- Omental or mesenteric metastases
- Fibrosing mesenteritis
- Hematoma
- Rhabdomyosarcoma
- Liposarcoma
- Gastrointestinal stromal tumors

enhancement of the most dominant mass extending down the inferior rectus abdominus muscle.

### **Differential diagnosis -- mesenteric masses**

- Desmoid tumor
- Carcinoid tumor
- Lymphoma
- Omental or mesenteric metastases
- Fibrosing mesenteritis
- Hematoma
- Rhabdomyosarcoma
- Liposarcoma
- Gastrointestinal stromal tumors

**Diagnosis:** Desmoid tumors

## Key points

### Desmoid tumors

- Desmoid tumors are benign proliferations of fibroblastic cells in connective or fibrous tissue.
- They are most commonly found in the abdomen, arising from the anterior abdominal wall, mesentery, or retroperitoneum.
- They are locally aggressive and have a high recurrence rate.
- Pathology:
  - 75% of patients have had prior abdominal surgery.
  - 18% to 20% of patients with Gardner's syndrome have it. Patients have a mutation in the APC gene.
  - Can be resistant to percutaneous biopsy because very hard.
  - Sporadically occurring tumors are more numerous than those seen in familial



- 75% of patients have had prior abdominal surgery.
- 18% to 20% of patients with Gardner's syndrome have it. Patients have a mutation in the APC gene.
- Can be resistant to percutaneous biopsy because very hard.
- Sporadically occurring tumors are more numerous than those seen in familial adenomatous polyposis (FAP) .
- The exact cause is unknown.
- Best diagnostic clue is a small-bowel mesentery or abdominal wall mass arising from a scar of prior surgery.
- Location:
  - Small-bowel mesentery, abdominal wall musculature, retroperitoneum, pelvic musculature
  - Extraabdominal locations: bladder, ribs, pelvic bones
- Range in size from 5 cm to 20 cm.
- Can be solitary or multiple.

## **Radiologic overview of the diagnosis**

- Ultrasonography: Appear with variable echogenicity, with smooth, well-defined margins.
- CT: May enhance and have either ill- or well- defined margins. Encase or displace mesenteric vessels.
- MRI: Low signal intensity relative to muscle on T1-weighted images and variable signal intensity on T2-weighted images. There are no specific imaging features to distinguish desmoid tumors from other solid masses.

## **Management and treatment**

- Wide surgical resection
- Radiation therapy for abdominal wall
- Steroids, nonsteroidal anti-inflammatory drugs (NSAIDs), chemotherapy, among other medications
- May require small-bowel transplant



- Radiation therapy for abdominal wall
- Steroids, nonsteroidal anti-inflammatory drugs (NSAIDs), chemotherapy, among other medications
- May require small-bowel transplant
- Recurrence rates of 25% to 65%

## Key points

- No specific imaging features to distinguish desmoid tumors from other solid masses.
- Most often seen in abdominal wall and small-bowel mesentery.
- Higher prevalence in patients with Gardner's syndrome.
- Has a high recurrence rate.
- The tumors are benign and rarely metastasize.
- Can cause complications from mass effect.
- Most effective treatment of accessible and smaller desmoid tumors is surgical resection with negative margins, although it may not prevent local recurrence.



# 48-year-old woman with incidental finding

CASE OUTLINE

Page 1 of 5

## History and PET/CT images

Our appreciation is extended to Dr. Teresa Martin-Carreras, University of Pennsylvania Department of Radiology, for contributing this case.

**History:** A 48-year-old woman with a remote history of melanoma was found to have a concerning lesion at C7 on a cervical spine MRI scan. She subsequently underwent a PET scan for further characterization of this lesion.

Subsequent imaging showed a hypermetabolic lesion at C7 on PET/CT scan. The PET/CT scan showed a hypermetabolic lesion at C7, which was consistent with the findings on the MRI scan.



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**Where is the highest uptake located?**

☐ Thyroid gland

☐ Tracheoesophageal groove

☐ Trachea

☐ Esophagus





**Where is the highest uptake located?**

☒ Thyroid gland (correct!)

☐ Tracheoesophageal groove

☐ Trachea

☐ Esophagus

☐ Esophagus

The question above accounts for 14% of your total score for this case.

**Each annihilation reaction in PET imaging results in the formation of two 511-keV energy photons that travel in opposite directions.**

☐ True

☐ False

The question above accounts for 16% of your total score for this case.

[VIEW YOUR SCORE](#)

☐ Esophagus

The question above accounts for 14% of your total score for this case.

**Each annihilation reaction in PET imaging results in the formation of two 511-keV energy photons that travel in opposite directions.**

☒ True (correct!)

☐ False

The question above accounts for 16% of your total score for this case.

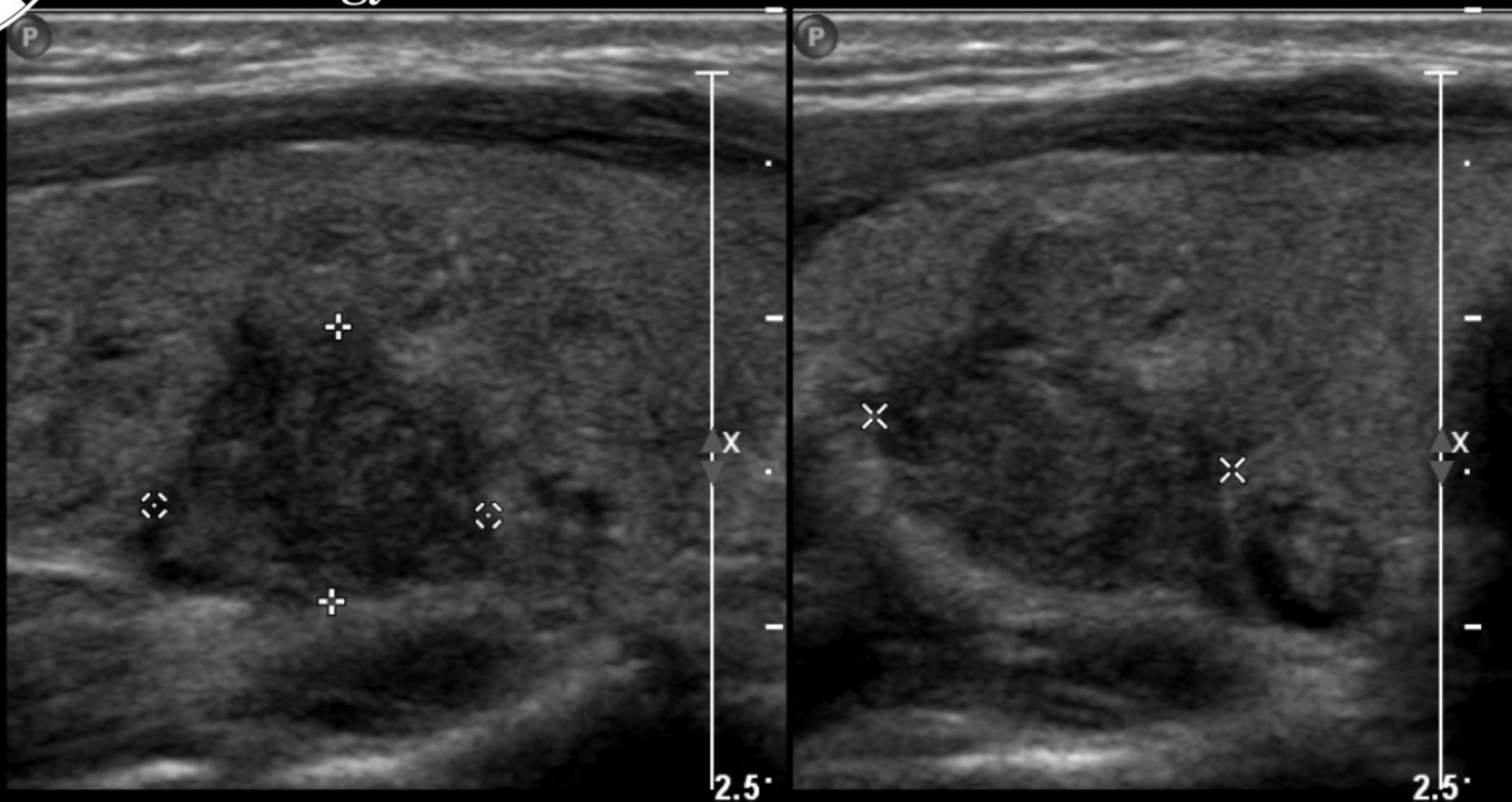
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# Penn

Radiology

OP Low  
Res



+ R Thyroid N1 AP 8.9 mm

x R Thyroid N1 Trv 11.7 mm

o R Thyroid N1 Long 10.8 mm

RIGHT THYROID N1 MID TRV



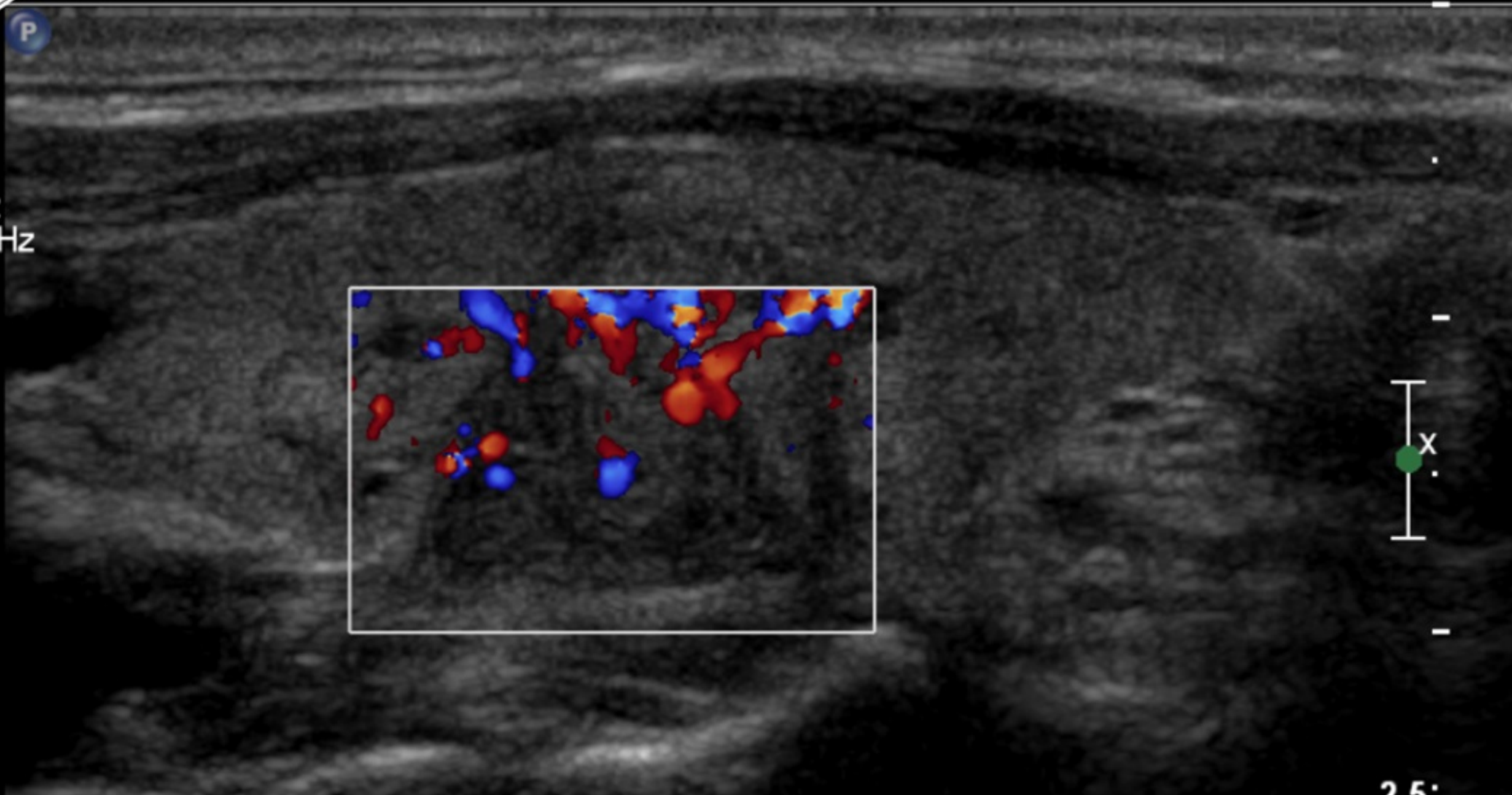


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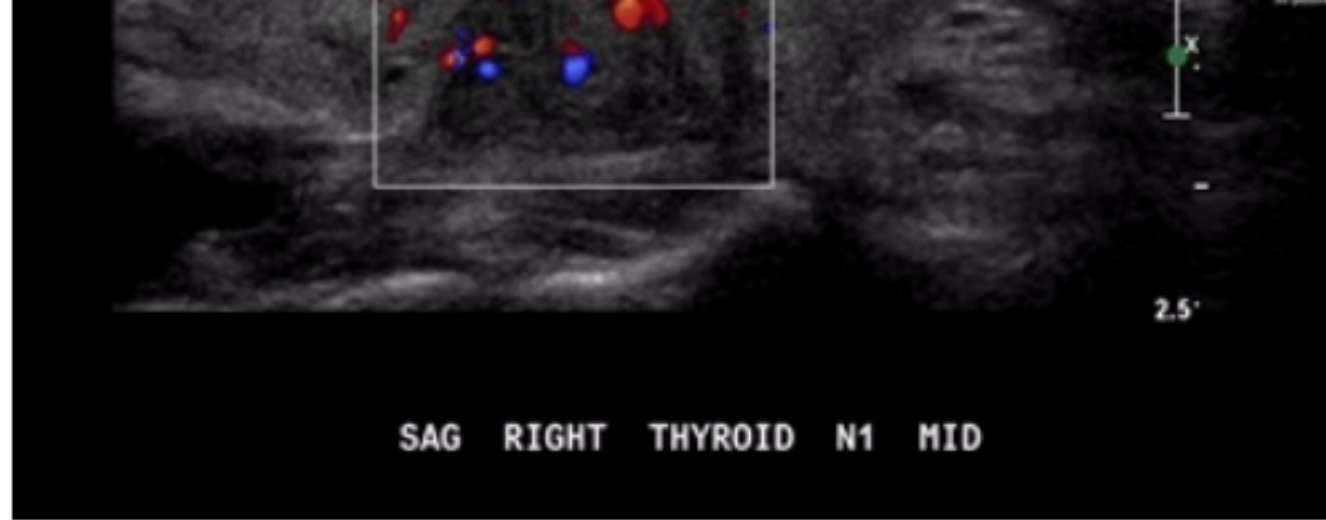
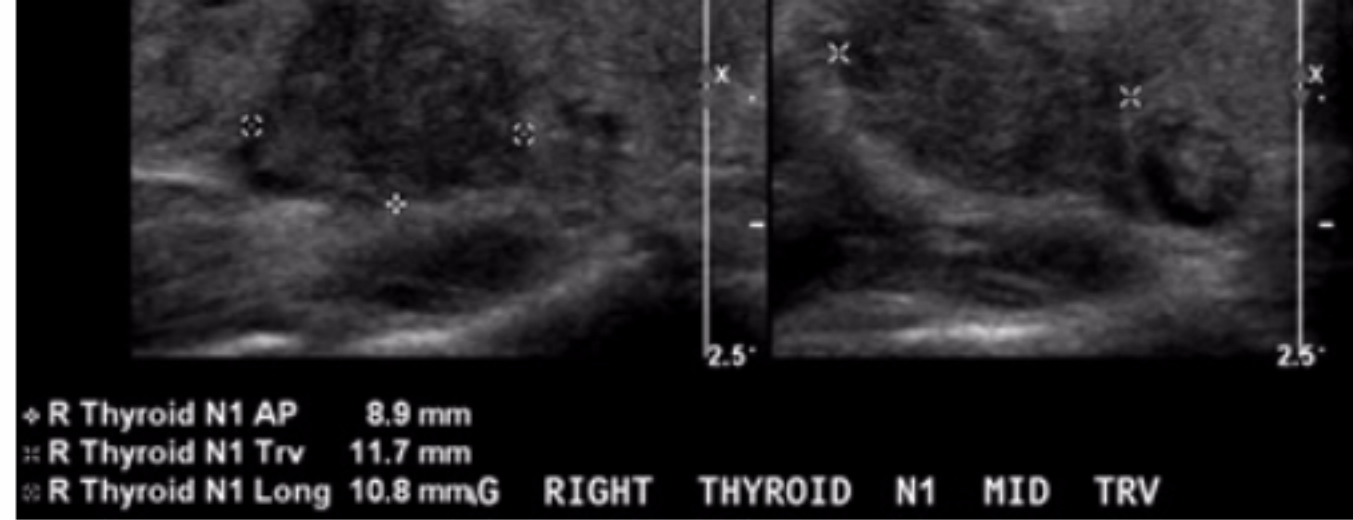
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80%  
700Hz  
WF 38Hz  
Low



2.5

SAG RIGHT THYROID N1 MID



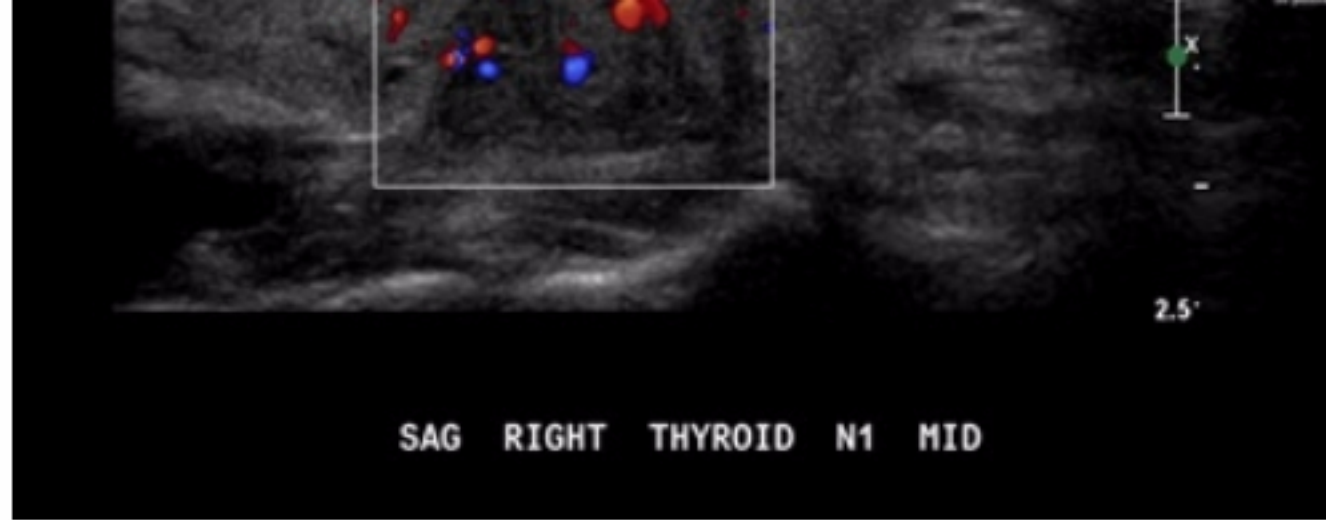


**Extensive calcification is noted within the thyroid gland nodule shown.**

☐ True

☐ False

The question above accounts for 16% of your total score for this case.



**Extensive calcification is noted within the thyroid gland nodule shown.**

☐ True

☒ False (correct!)

The question above accounts for 16% of your total score for this case.

The question above accounts for 16% of your total score for this case.

**Fine-needle aspiration (FNA) of the thyroid nodule is the recommended next step in management.**

☐ True

☐ False

The question above accounts for 16% of your total score for this case.

[VIEW YOUR SCORE](#)

The question above accounts for 16% of your total score for this case.

**Fine-needle aspiration (FNA) of the thyroid nodule is the recommended next step in management.**

☒ True (correct!)

☐ False

The question above accounts for 16% of your total score for this case.

[VIEW YOUR SCORE](#)



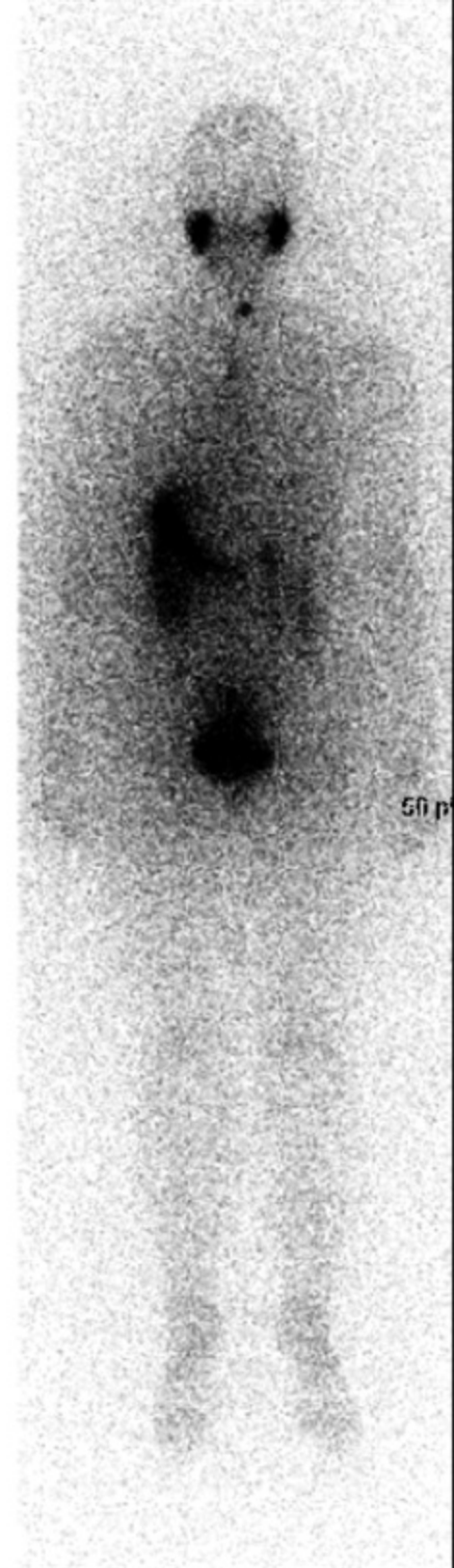
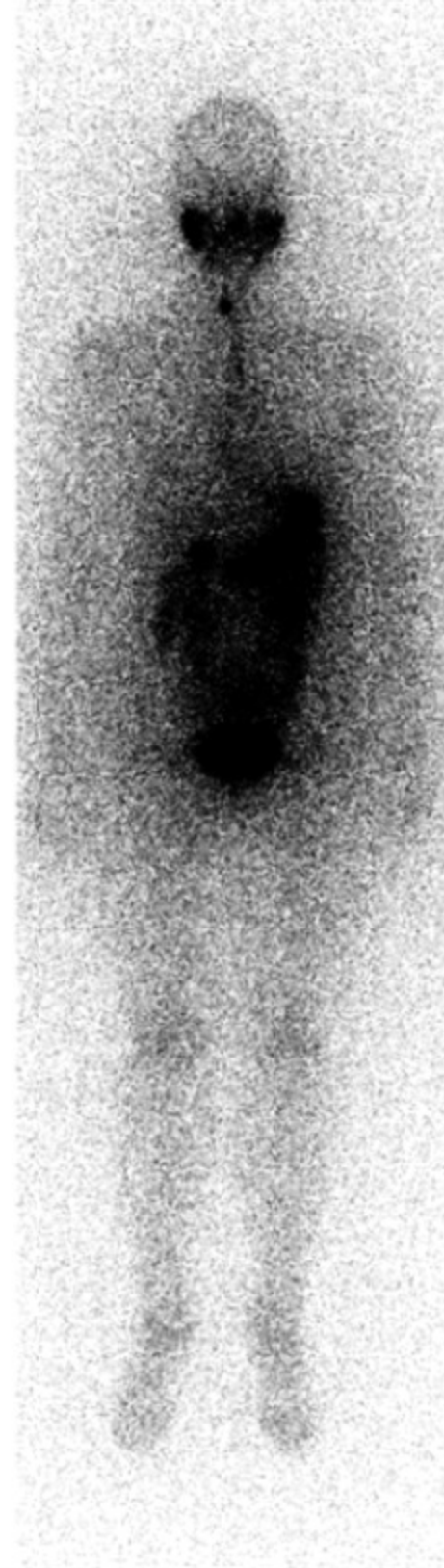
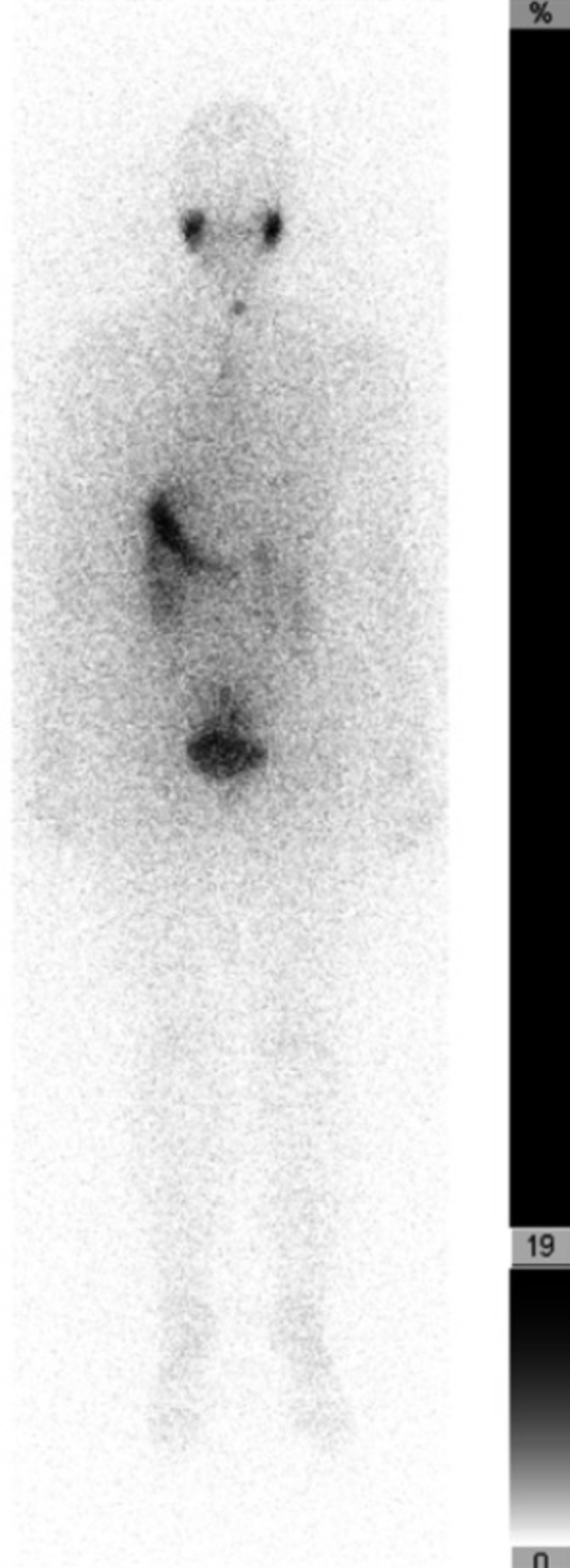
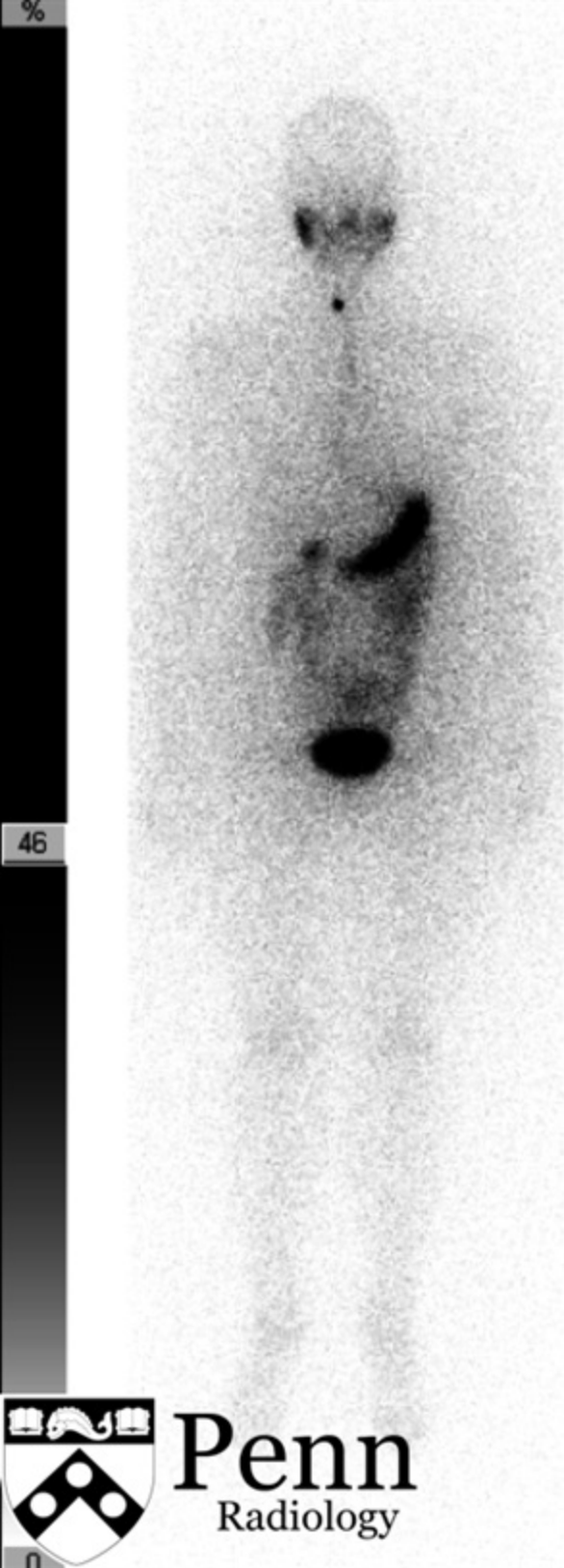
## Scintigraphy images

The patient underwent a total thyroidectomy with pathology consistent with papillary thyroid cancer (tall cell variant), and she presented several months later to be evaluated for treatment with radioactive iodine (RAI) ablation therapy. The patient was pretreated with two daily intramuscular doses of 0.9-mg recombinant human thyroid-stimulating hormone (rhTSH) and followed a low-iodine diet, and all laboratory values were within acceptable ranges.

Thyroid scintigraphy with iodine-123 (I-123) was subsequently performed. Anterior and posterior whole-body images were acquired approximately 24 hours later and were supplemented by spot images of the neck. Click images below to enlarge.



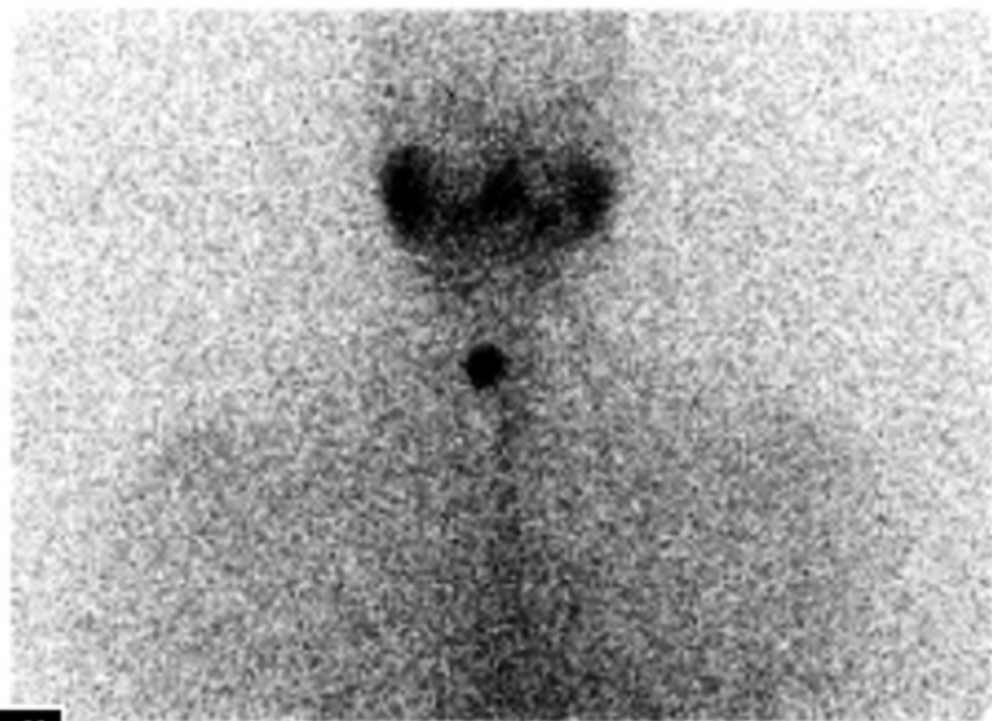




**Penn**  
Radiology

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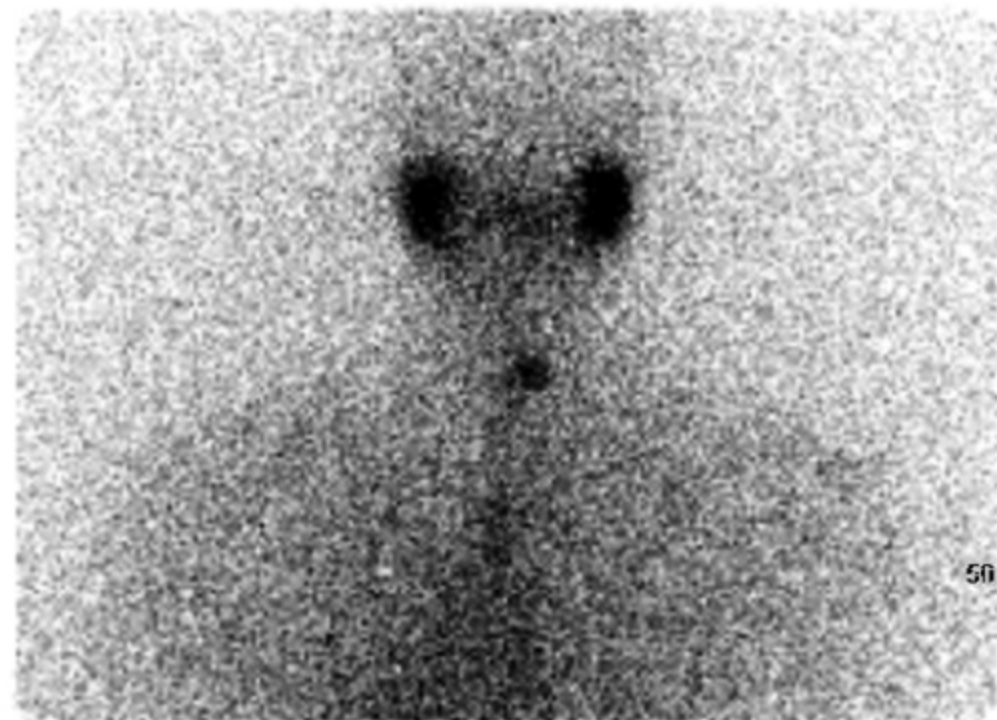
52



**Penn**  
Radiology

%

53



50 pixels

The patient was subsequently treated with I-131 RAI ablation therapy.

**There are distant radioiodine-avid lesions suggestive of distant metastases.**

☐ True

☐ False

The question above accounts for 16% of your total score for this case.

**Which of the following corresponds to the photon energy and physical half**



The patient was subsequently treated with I-131 RAI ablation therapy.

**There are distant radioiodine-avid lesions suggestive of distant metastases.**

☐ True

☒ False (correct!)

The question above accounts for 16% of your total score for this case.

**Which of the following corresponds to the photon energy and physical half**

The question above accounts for 16% of your total score for this case.

**Which of the following corresponds to the photon energy and physical half-life of I-123, respectively?**

☐ 364 keV, 8 days

☐ 159 keV, 13.2 hours

☐ 81 keV, 5.27 days

☐ 80 keV, 55 hours



The question above accounts for 16% of your total score for this case.

**Which of the following corresponds to the photon energy and physical half-life of I-123, respectively?**

☐ 364 keV, 8 days

☒ 159 keV, 13.2 hours (correct!)

☐ 81 keV, 5.27 days

☐ 80 keV, 55 hours

## Findings

- **PET/CT:** No FDG-avid focus is noted in the C7 vertebral body to correspond with the lesion on MRI. There is a 7-mm FDG-avid nodule in the right thyroid gland with maximum standard uptake value (SUV) of 15.99.
- **Ultrasound:** There is a noncalcified, hypoechoic, infiltrative-appearing solid nodule in the posterior mid right thyroid lobe, measuring 9 x 12 x 11 mm. This nodule corresponds to the FDG-avid nodule seen on PET/CT. Fine-needle aspiration biopsy of the mid right posterior thyroid lobe nodule (N1) is recommended based on its size, appearance, and FDG avidity. This is favored to represent a nodule of thyroid origin, as opposed to a metastatic lesion.
- **I-123 thyroid scan:** In the region of the right thyroidectomy bed, there is focal radiotracer uptake consistent with residual tissue of thyroidal origin. There are no radioiodine avid lesions suggestive of distant metastases. Physiologic tracer

FDG avidity. This is favored to represent a nodule of thyroid origin, as opposed to a metastatic lesion.

- **I-123 thyroid scan:** In the region of the right thyroidectomy bed, there is focal radiotracer uptake consistent with residual tissue of thyroidal origin. There are no radioiodine avid lesions suggestive of distant metastases. Physiologic tracer distribution is visualized in the salivary glands, gastrointestinal tract, and urinary bladder. Quantification of uptake in the neck was 1.5% at 21 hours.

## Differential diagnosis

- Papillary thyroid cancer
- Follicular thyroid cancer
- Medullary thyroid cancer
- Anaplastic thyroid cancer

**Diagnosis:** Papillary thyroid cancer, tall cell variant



# Papillary thyroid cancer (PTC)

## Pathophysiology

- Papillary thyroid cancer is microscopically characterized by the presence of papillae consisting of one or two layers of tumor cells surrounding a fibrovascular stalk.
- About 50% contain calcified psammoma bodies, scarred remnants of tumor papillae that have likely infarcted.
- PTC is typically unencapsulated, and lymph node involvement is very common.
- PTC is radioactive iodine (RAI)-avid and often multifocal within the gland.
- Variant forms of PTC include follicular, tall cell, insular, columnar, solid, clear cell, diffuse sclerosing, cribriform morular, Hürthle cell, and hobnail variants.
- Mutations or rearrangements in the genes encoding for the proteins in the mitogen-activated protein kinase (MAPK) pathway are important to the development and progression of differentiated PTC. Mutations in RET/PTC, NTRK1, Ras, or BRAF occur in as many as 70% of well-differentiated PTCs.

- Prognosis is very good with a five-year survival of approximately 95%. Poor prognostic indicators include tumors larger than 1.5 cm, extracapsular invasion, and age older than 45 years.
- Approximately 2% to 10% of patients have metastatic disease beyond the neck at the time of diagnosis.
  - Two-thirds of patients have pulmonary metastases, and one-fourth have skeletal metastases.
  - Rare sites of metastasis include the brain, kidneys, liver, and adrenals.

## Epidemiology

- Peak incidence for papillary thyroid cancer is in the third and fourth decades
- It is more common in women (male:female ratio of 1:1.6-3.1).
- PTC accounts for approximately 70% of thyroid neoplasms and 85% of thyroid cancers.
- Risk factors include previous head/neck radiation, a family history of thyroid cancer, age younger than 20 or older than 50 years, and female gender.



- Risk factors include previous head/neck radiation, a family history of thyroid cancer, age younger than 20 or older than 50 years, and female gender.
- In the U.S., there are approximately 62,980 cases of thyroid cancer and about 1,890 deaths from thyroid cancer per year.
- Papillary thyroid cancer has the fastest increasing incidence of any malignancy in the thyroid.

## **Clinical presentation**

- Patients most commonly present with a painless, solitary palpable thyroid mass.
- Approximately 50% of patients may present with hoarseness due to local invasion.
- If the disease is advanced at the time of presentation, patients may have a fixed hard mass, lymphadenopathy, and/or vocal cord paresis at the time of presentation.

## **Imaging features**

L422 whole body scan

## Imaging features

- I-123 whole-body scan:
  - I-123 localizes to thyroid tissue that transports and organifies iodine.
  - Detects thyroid remnant, lymphadenopathy, and distant metastatic or recurrent disease.
  - Physiologic uptake is seen in the salivary glands, stomach, lactating breasts.
  - Excretion is renal.
  - 159 keV gamma rays are more optimal for low-energy all-purpose (LEAP) collimators.
  - Patients need an elevated thyroid-stimulating hormone (TSH), which can be achieved via thyroid hormone withdrawal for three weeks (goal TSH  $> 30$  mU/L) or Thyrogen (rhTSH) stimulation with 0.9-mg intramuscular doses on two consecutive days.
  - Patients should also maintain a low-iodine diet for seven to 14 days to increase uptake of radioactive iodine by thyroid tissue.



uptake of radioactive iodine by thyroid tissue.

- I-131 whole-body scan:
  - I-131 is a gamma (364 keV) and beta (0.606 MeV) emitter.
  - There is a potential risk of thyroid stunning prior to I-131 ablation due to high-energy beta.
  - It has a higher radiation burden as compared to I-123.
  - Imaging characteristics are not as good as I-123.
- F-18 FDG-PET/CT:
  - Uptake of radioactive glucose analogue into thyroid tissue is via glucose transporter (GLUT1).
  - GLUT1 has been found to be overexpressed in thyroid carcinomas, thus increased uptake of tracer suggests the possibility of carcinoma.
  - A solitary thyroid nodule with increased FDG activity has approximately a 1 in 3 chance of being malignant.
  - Tumors that are radioiodine-negative and F-18 FDG-positive often have a less favorable prognosis.

favorable prognosis.

- Technetium-99m methyl diphosphonate (Tc-99m MDP) bone scan: Can be used for a skeletal survey if osseous metastatic disease is suspected.
- Ultrasound:
  - Approximately 10% to 20% of PTCs are multifocal, 70% are solid, and 77% to 90% are hypoechoic.
  - Commonly have irregular and ill-defined margins.
  - Large tumors may invade strap muscles, esophagus, trachea, recurrent laryngeal nerve, and neck vessels.
  - Color Doppler: Chaotic vascularity within the nodule, wall, and septa in nodules with cystic change.
  - Nodes are predominantly hyperechoic (80%) compared with muscles, and approximately 50% have punctate microcalcifications.

## Treatment

Treatment commonly entails a total thyroidectomy with or without lymphadenectomy for



- Nodes are predominantly hyperechoic (80%) compared with muscles, and approximately 50% have punctate microcalcifications.

## Treatment

- Treatment commonly entails a total thyroidectomy with or without lymphadenectomy for clinical nodal disease.
- Postoperative radioactive iodine (RAI) ablation therapy also can be part of treatment as per American Thyroid Association guidelines.
- Screening after surgery is performed with ultrasound plus serum thyroglobulin levels for assessment of recurrence.
- If clinical suspicion of recurrence is high, a whole-body radioactive iodine scan may be performed.
- If there is disease progression despite RAI, brain MRI, neck contrast-enhanced CT, and chest high-resolution CT should be considered.



## History and mammography images

Our appreciation is extended to Dr. Erica Alexander, University of Pennsylvania Department of Radiology, for contributing this case.

**History:** An 89-year old woman presents with left-sided breast pain and a palpable lump for approximately one month.

A diagnostic mammogram was ordered. Left and right craniocaudal (CC) and mediolateral (MLO) views are shown below. Click to enlarge.





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RCC



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Radiology



**Penn**  
Radiology



RMLO



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Radiology



## Where is the breast abnormality located?

☐ Left upper outer

☐ Left upper inner

☐ Left central

☐ Left lower outer

☐ Left lower inner

## Where is the breast abnormality located?

☐ Left upper outer

☐ Left upper inner

☒ Left central (correct!)

☐ Left lower outer

☐ Left lower inner

The question above accounts for 15% of your total score for this case.

**Which of the following best describes the breast density using the BI-RADS reporting system?**

☐ Fatty

☐ Scattered

☐ Heterogeneous

☐ Dense

The question above accounts for 15% of your total score for this case.

**Which of the following best describes the breast density using the BI-RADS reporting system?**

☐ Fatty

☒ Scattered (correct!)

☐ Heterogeneous

☐ Dense

**What is the next most appropriate step in the management of this patient?**

☐ Ultrasound

☐ MRI

☐ Biopsy

☐ Follow-up in three months to assess stability

The question above accounts for 15% of your total score for this case.



**What is the next most appropriate step in the management of this patient?**

☒ Ultrasound (correct!)

☐ MRI

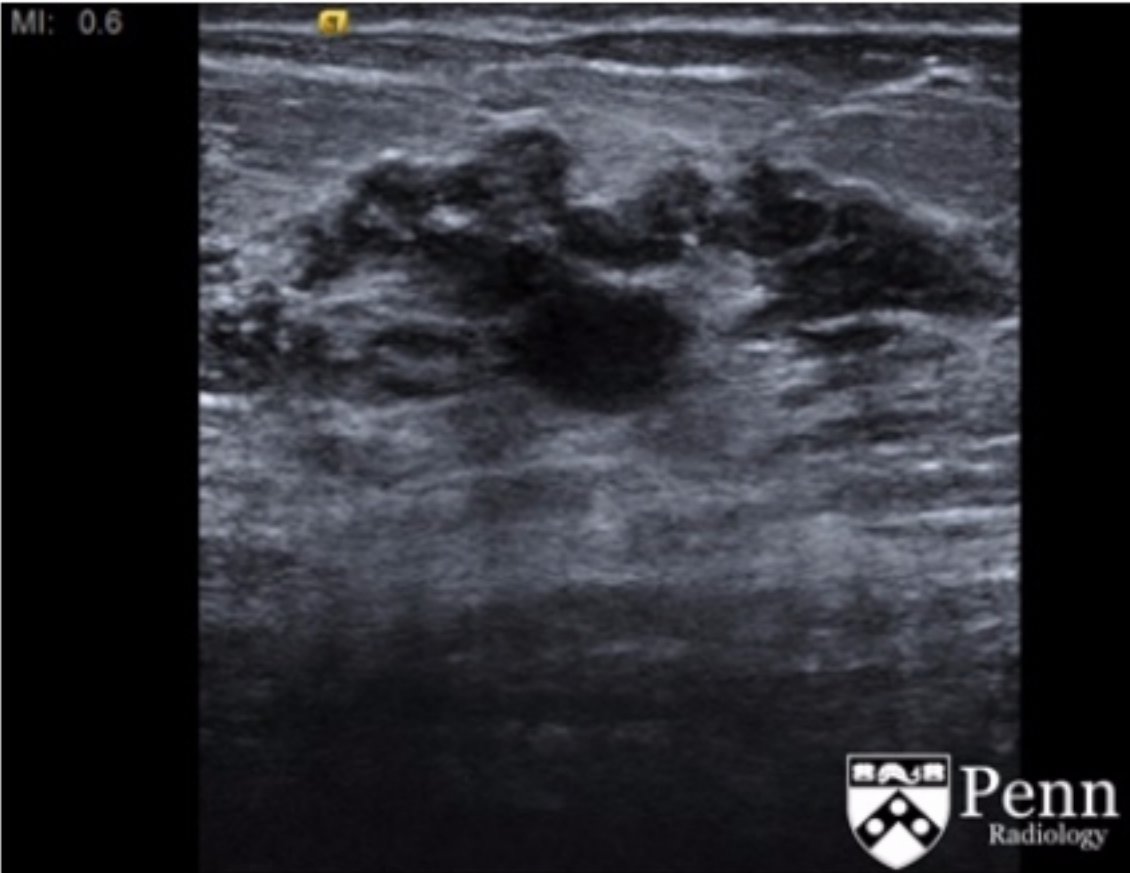
☐ Biopsy

☐ Follow-up in three months to assess stability

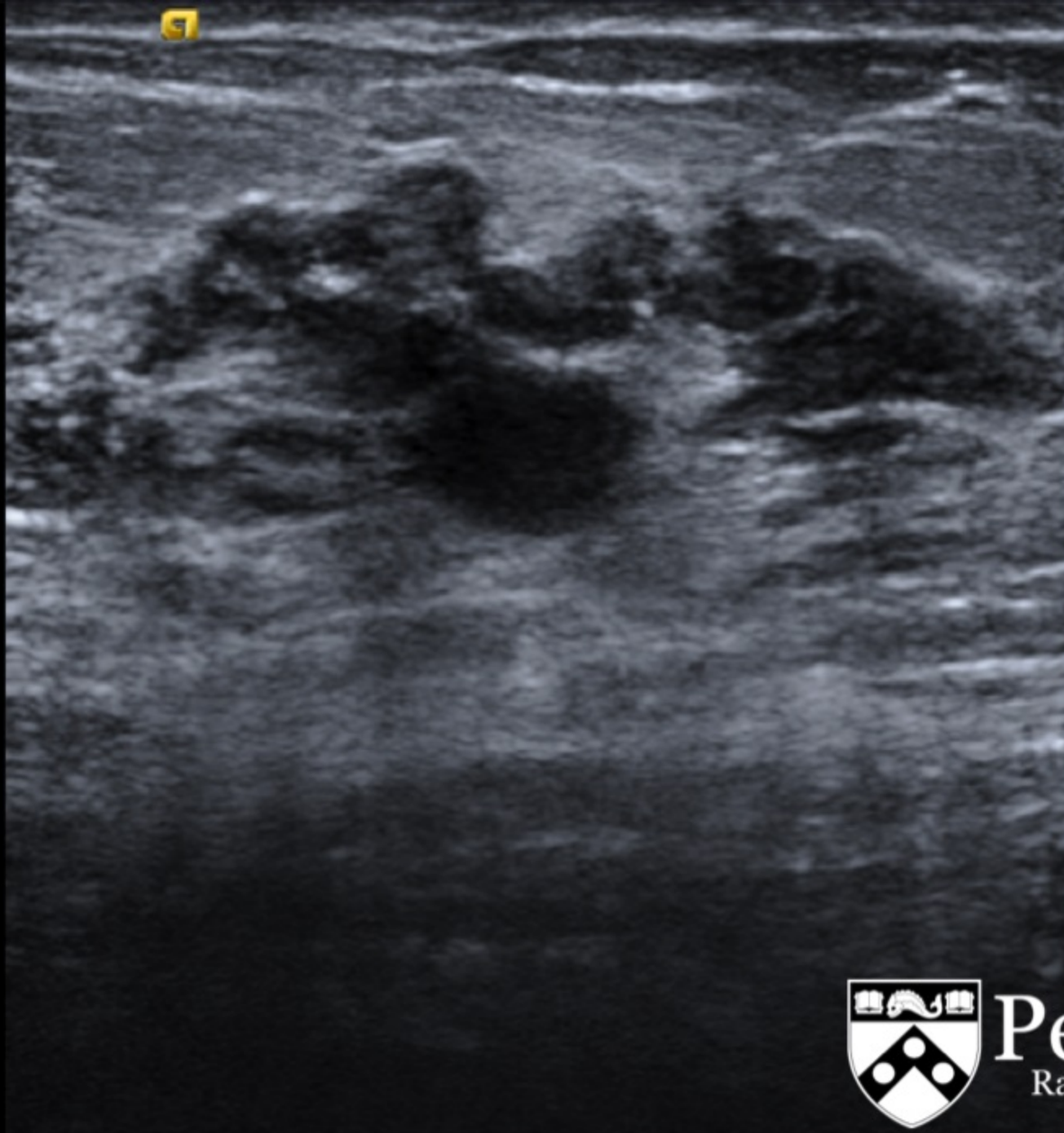
The question above accounts for 15% of your total score for this case.

# Ultrasound images

Targeted ultrasound images of the left breast at 12 to 1 o'clock are shown below. Click to enlarge.



MI: 0.6



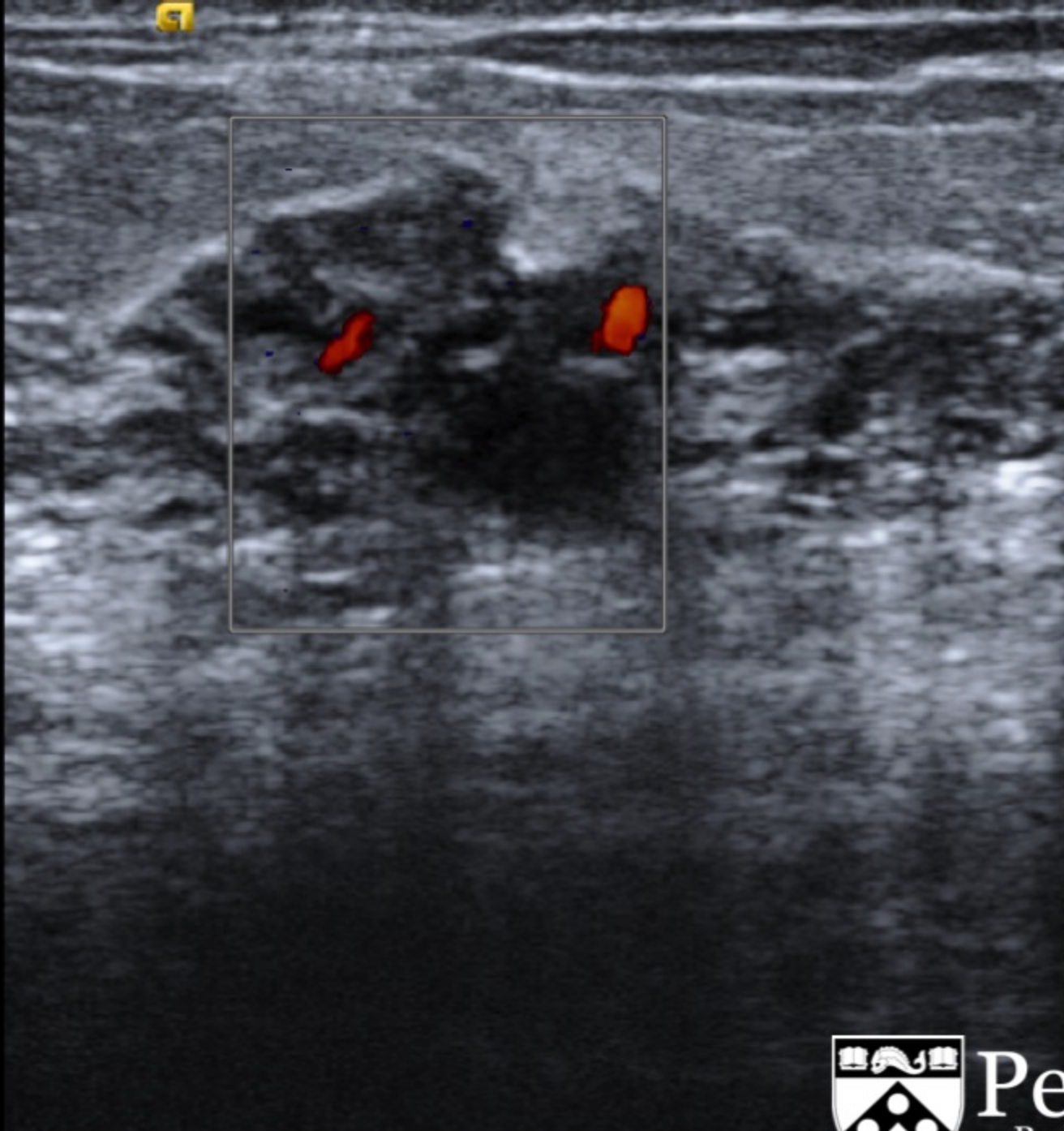
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LEFT BREAST 12-100 3 CM FAM ARAD \_

60fps

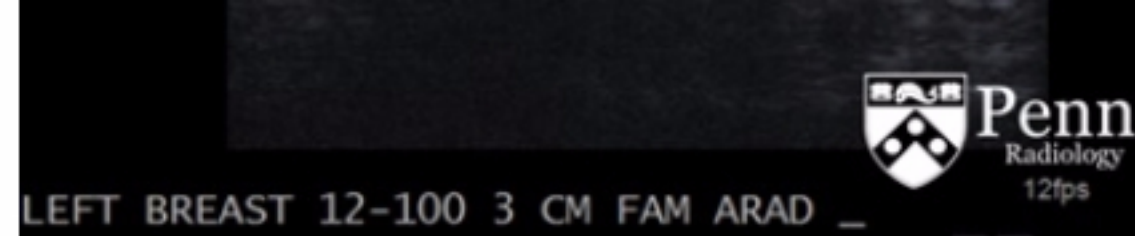
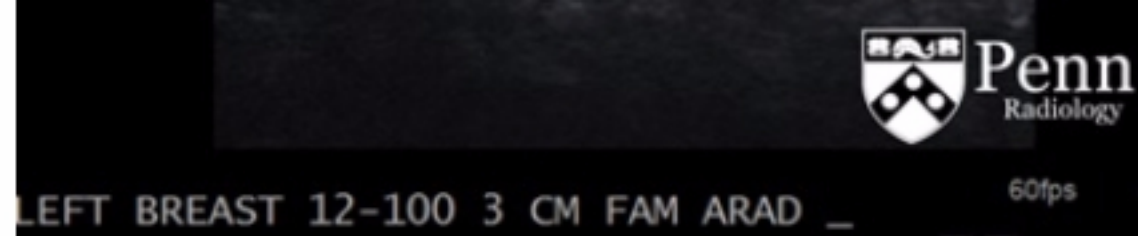


TIS: 1.2  
TIB: 1.2  
MI: 1.4



Penn  
Radiology  
12fps

LEFT BREAST 12-100 3 CM FAM ARAD \_



**The ultrasound reveals echogenic cysts in the left central breast.**

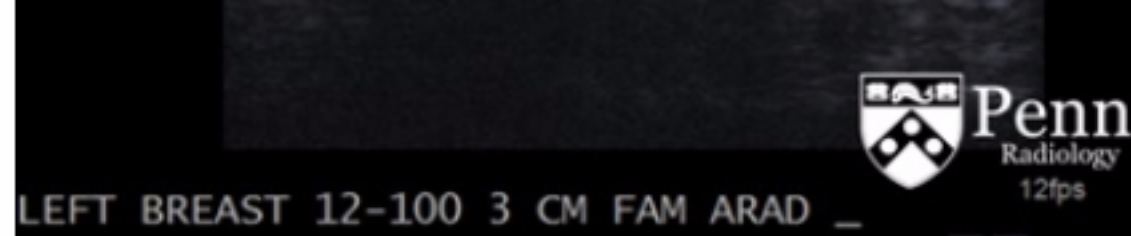
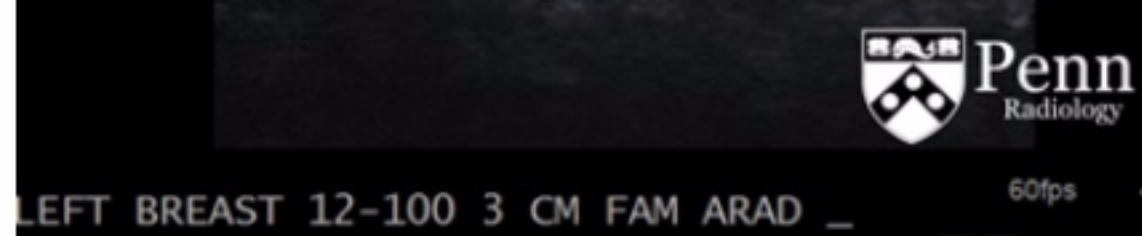
☐ True

☐ False

The question above accounts for 15% of your total score for this case.

**What BI-RADS score is most appropriate for this patient?**





**The ultrasound reveals echogenic cysts in the left central breast.**

☐ True

☒ False (correct!)

The question above accounts for 15% of your total score for this case.

**What BI-RADS score is most appropriate for this patient?**

The question above accounts for 15% of your total score for this case.

**What BI-RADS score is most appropriate for this patient?**

☐ BI-RADS 0

☐ BI-RADS 3

☐ BI-RADS 4

☐ BI-RADS 5

The question above accounts for 20% of your total score for this case.

The question above accounts for 15% of your total score for this case.

**What BI-RADS score is most appropriate for this patient?**

☐ BI-RADS 0

☐ BI-RADS 3

☒ BI-RADS 4 (correct!)

☐ BI-RADS 5

The question above accounts for 20% of your total score for this case.

The question above accounts for 20% of your total score for this case.

**What is now the most appropriate stage in management?**

☐ MRI

☐ Biopsy

☐ Follow-up ultrasound in three months

☐ Diagnostic mammogram in six months

The question above accounts for 20% of your total score for this case

The question above accounts for 20% of your total score for this case.

**What is now the most appropriate stage in management?**

☐ MRI

☒ Biopsy (correct!)

☐ Follow-up ultrasound in three months

☐ Diagnostic mammogram in six months

The question above accounts for 20% of your total score for this case



## Findings

- **Mammography:** There are scattered fibroglandular densities in the bilateral breasts. There is an area of asymmetry in the central left breast, in the area of palpable concern. Images of the right breast do not demonstrate any suspicious findings.
- **Ultrasound:** Targeted ultrasound of the left central breast reveals an ill-defined mass measuring 3.5 cm in the 12 to 1 o'clock region. Color Doppler reveals internal vascularity.

## Differential diagnosis

- Invasive ductal carcinoma
- Lymphoma
- Metastatic disease
- Phyllodes tumor

vascularity.

## Differential diagnosis

- Invasive ductal carcinoma
- Lymphoma
- Metastatic disease
- Phyllodes tumor
- Fibroadenoma
- Galactocele
- Abscess
- Complicated cyst

**Diagnosis:** The overall BI-RADS score was category 4, suspicious. The patient went on to have a breast biopsy, and pathology revealed intraductal carcinoma, papillary type.

# Keypoints

## Papillary breast cancer

### Pathophysiology

Papillary breast cancers begin in the milk ducts of the breast. Papillary breast cancers tend to be small and estrogen receptor-positive and progesterone receptor-positive (ER/PR-positive) and HER2 receptor-negative.

They include a heterogeneous group of breast lesions that share a common growth pattern of arborescent fibrovascular stalks lined with epithelial cells. Malignant papillary neoplasms include a number of microscopically distinct lesions, including ductal carcinoma in situ (DCIS) arising from an intraductal papilloma, solid papillary carcinoma, encapsulated papillary carcinoma, and invasive papillary carcinoma. Malignant papillary carcinomas are distinguished from benign intraductal papillomas in that they lack an intact myoepithelial cell



papillary carcinoma, and invasive papillary carcinoma. Malignant papillary carcinomas are distinguished from benign intraductal papillomas in that they lack an intact myoepithelial cell layer within the papillae.

Histology suggests that papillary breast cancer is caused by a proliferation of cells arranged around fibrovascular cores, which grossly form a circumscribed mass.

## **Epidemiology**

Papillary breast cancers are quite rare and account for approximately 0.5% of invasive breast cancers that are diagnosed. They tend to affect postmenopausal women ages 65 to 70. They have a relatively good prognosis.

## **Clinical presentation**

Patients typically present with bloody nipple discharge and/or an abnormal mass. The masses may be large secondary to the cystic component of the mass.

## Imaging features

- Ultrasound: Papillary neoplasms present with the following sonographic findings: an intraductal mass with or without ductal dilatation, an intracystic mass, and/or a solid intraductal mass completely filling the duct. They tend to have papillary projections and fronds. Hemorrhage may be seen on ultrasound. On color Doppler, these masses tend to display increased vascularity or a large feeding vessel.
- Mammography: Papillary masses tend to be round, oval, or lobulated in shape. They can have partially circumscribed and partially indistinct margins, along with amorphous or pleomorphic calcifications.
- MRI: On enhanced T1-weighted images, papillary neoplasms tend to present as an intracystic mass with irregular, enhancing mural nodules. Hemorrhage may or may not be seen. There is heterogeneous internal enhancement.



Intracystic mass with irregular, enhancing mural nodules. Hemorrhage may or may not be seen. There is heterogeneous internal enhancement.

## Treatment

Treatment depends on the stage of the tumor, tumor features, and the overall health of the patient.

Local therapy includes surgery with lymph node sampling and/or radiation. Systemic therapy, such as endocrine hormone therapy, chemotherapy, and targeted HER2 monoclonal antibodies, can be considered.

## References

1. Collins LC, Schnitt SJ. Papillary lesions of the breast: Selected diagnostic and management issues. *Histopathology*. 2008;52(1):20-29.
2. Fiada B, Cheng J, Kulkarni S, Goldberg E, Muradali D. Papillary lesions of the breast:

## History and radiographs

Our appreciation is extended to Dr. Akash Patel, University of Pennsylvania Department of Radiology, for contributing this case.

**History:** A 19-year-old man presents with progressive, right-sided hip pain.

Frontal radiographs of the pelvis and right hip are shown below. Click to enlarge.

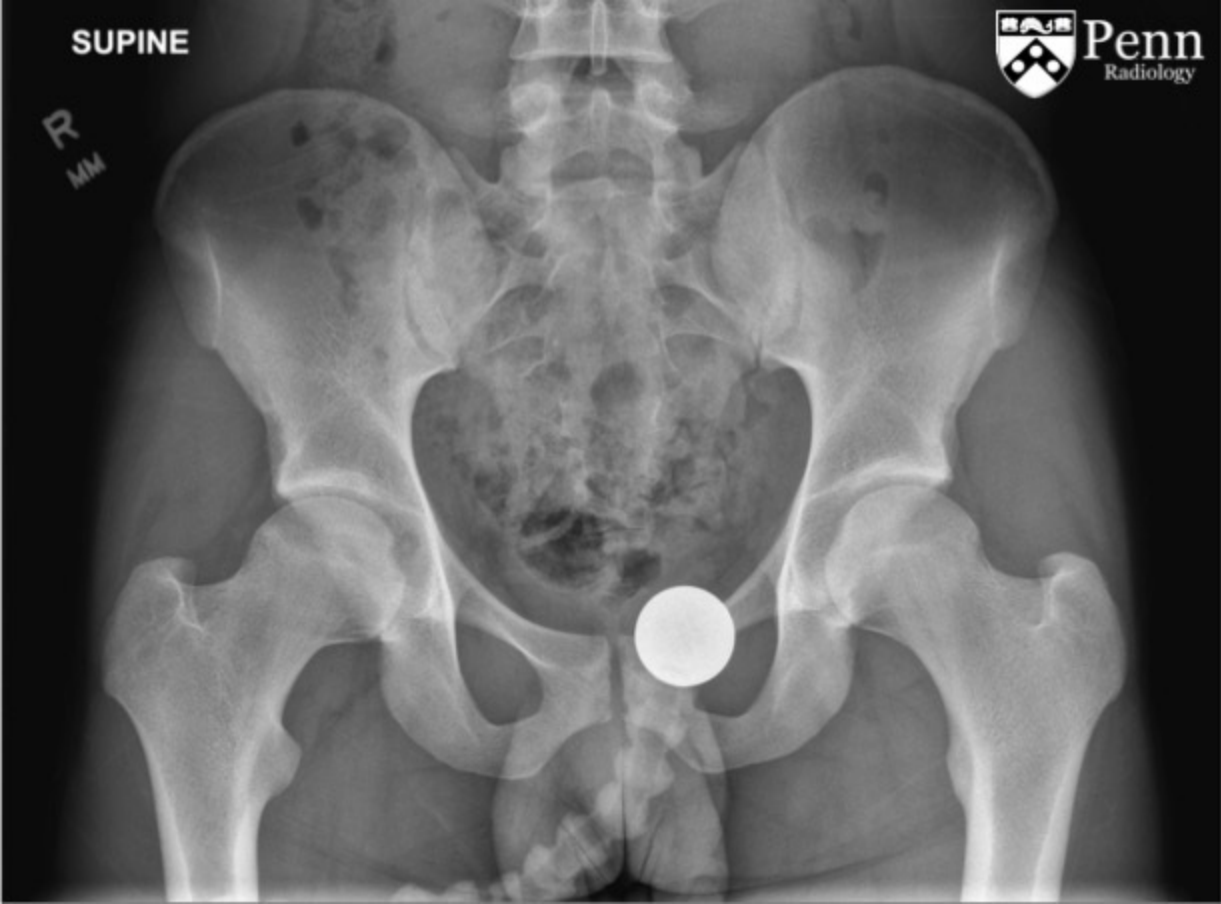


SUPINE



Penn  
Radiology

R  
MM





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Radiology



## Where is the abnormality?

- ☐ Spine
- ☒ Right acetabulum
- ☐ Right femur
- ☐ None of the above

The question above accounts for 16% of your total score for this case.



## Where is the abnormality?

☐ Spine

☒ Right acetabulum (correct!)

☐ Right femur

☐ None of the above

**[Explain this Answer]**

The question above accounts for 16% of your total score for this case

**[Explain this Answer]**

The question above accounts for 16% of your total score for this case.

**What radiographic feature does this lesion demonstrate?**

☐ Lucent with central focus of increased density

☐ Uniform density

☐ Involves multiple bones

The question above accounts for 16% of your total score for this case.

**[Explain this Answer]**

The question above accounts for 16% of your total score for this case.

**What radiographic feature does this lesion demonstrate?**

☒ Lucent with central focus of increased density (correct!)

☐ Uniform density

☐ Involves multiple bones

**[Explain this Answer]**

The question above accounts for 16% of your total score for this case.

**What is the next best study to characterize the suspected lesion?**

☐ Bone scan

☐ Ultrasound

☐ CT

☐ PET/CT

The question above accounts for 17% of your total score for this case.

**What is the next best study to characterize the suspected lesion?**

☐ Bone scan

☐ Ultrasound

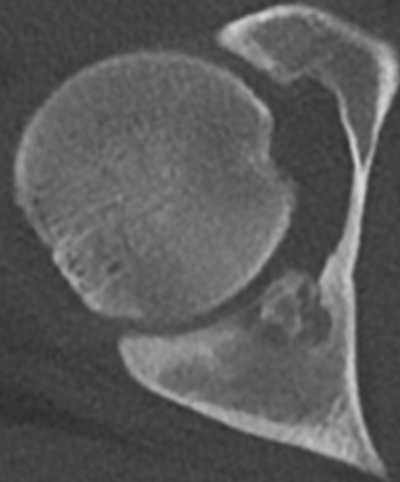
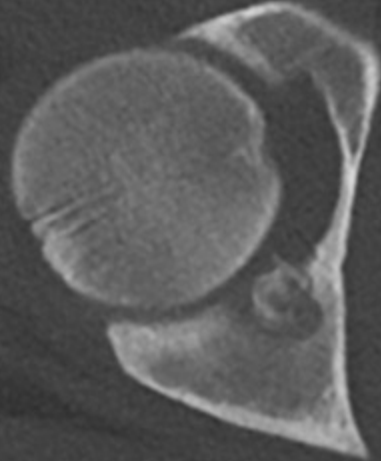
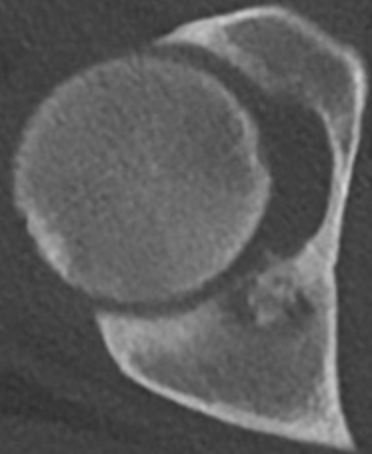
☒ CT (correct!)

☐ PET/CT

**[Explain this Answer]**

The question above accounts for 17% of your total score for this case.







**The lesion contains a central sclerotic nidus.**

☐ True

☐ False

The question above accounts for 17% of your total score for this case.

**The lesion is malignant in nature.**

☐ True

**The lesion contains a central sclerotic nidus.**

☒ True (correct!)

☐ False

The question above accounts for 17% of your total score for this case.

**The lesion is malignant in nature.**

☐ True

The question above accounts for 17% of your total score for this case.

**The lesion is malignant in nature.**

☐ True

☐ False

The question above accounts for 17% of your total score for this case.

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[2](#)

[3](#)

[5](#)

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The question above accounts for 17% of your total score for this case.

**The lesion is malignant in nature.**

☐ True

☒ False (correct!)

**[Explain this Answer]**

The question above accounts for 17% of your total score for this case.

[VIEW YOUR SCORE](#)

## Findings

- **Radiographs:** There is a small, round, radiolucent lesion of the right acetabulum with a central focus of increased density.
- **CT:** The radiolucent lesion is seen in the cortex of the right acetabulum with a central sclerotic nidus.

## Differential diagnosis

- Osteoid osteoma
- Osteoblastoma
- Chronic cortical osteomyelitis
- Osteosarcoma
- Ewing's sarcoma

**Diagnosis:** Osteoid osteoma

## Additional question

**Classically, patients with this lesion have symptomatic relief with which of the following medications?**

☐ Antibiotics

☒ Nonsteroidal anti-inflammatory drugs (NSAIDs)

☐ Diuretics

The question above accounts for 17% of your total score for this case.

## Additional question

**Classically, patients with this lesion have symptomatic relief with which of the following medications?**

☐ Antibiotics

☒ Nonsteroidal anti-inflammatory drugs (NSAIDs) (correct!)

☐ Diuretics

The question above accounts for 17% of your total score for this case.

# Osteoid osteoma

## Pathophysiology

- Osteoid osteoma is a solitary, benign bone lesion, usually occurring in a long bone cortex.
- Its pathogenesis is controversial.
  - Some believe it arises on an inflammatory basis.
  - Some suggest it is an unusual healing and reparative process.
- Characteristically and unlike other bone tumors, it is composed of a nidus with a relatively immature central area and more mature calcified peripheral bone.
- It has little to no growth potential.
- They rarely exceed 1.5 cm.

## Epidemiology



## Epidemiology

- Osteoid osteoma is usually found in the lower extremities of children and young adults, with the proximal femur as the most common location.
- Most commonly occurs in the second and third decades.
- It is the third most common benign bone tumor.
  - 10% to 11% of benign bone tumors
  - 3% of all primary bone tumors
- The male-to-female ratio is 3:1.

## Clinical presentation

- Most patients present with severe pain.
  - The pain is characteristically increased at night.
  - NSAIDs are used for pain relief.
  - Prostaglandin production is believed to be a major factor.

- Prostaglandin production is believed to be a major factor.
- Duration of pain varies from weeks to years.
- Osteoid osteomas are often misdiagnosed clinically.

## Imaging features

- Radiographs:
  - Classically, radiographs show an area of cortical thickening and sclerosis containing a lucent central nidus ( $< 1$  cm).
  - A nidus is present in 85% of cases.
  - 20% intramedullary with less surrounding sclerosis.
  - Subperiosteal location often has limited surrounding sclerotic response.
- CT:
  - Commonly considered to be the most valuable imaging modality for diagnosis and localization.
  - CT offers better visualization of the nidus.
- MRI: Cortically based lesions are less conspicuous on MRI due to decreased spatial

- CT offers better visualization of the nidus.
- MRI: Cortically based lesions are less conspicuous on MRI due to decreased spatial resolution and volume averaging.
- Bone scan -- technetium-99
  - Useful if osteoid osteoma is suspected but not obvious on radiographs.
  - Shows avid radiotracer uptake during vascular, blood-pool, and delayed phases.
  - Classic “double-density” appearance is very specific.

## Differential diagnoses

- Osteoblastoma -- some believe it is the same lesion at a different stage of development
- Chronic cortical osteomyelitis
- Osteosarcoma
- Ewing's sarcoma

## Treatment



## Treatment

- Conservative treatment:
  - NSAIDs are used for pain relief.
  - Osteoid osteomas may undergo spontaneous regression after several years.
- Percutaneous -- many options are available, including the following:
  - MRI-guided cryotherapy
  - CT-guided drilling of the nidus with ethanol injection
  - Radiofrequency thermocoagulation
- Surgery:
  - Osteoid osteomas are often difficult to identify.
  - Incomplete resection may lead to recurrence.

## References

1. Accoun J, Richardi C, Bailhe J, et al. Osteoid osteoma: MR imaging versus CT

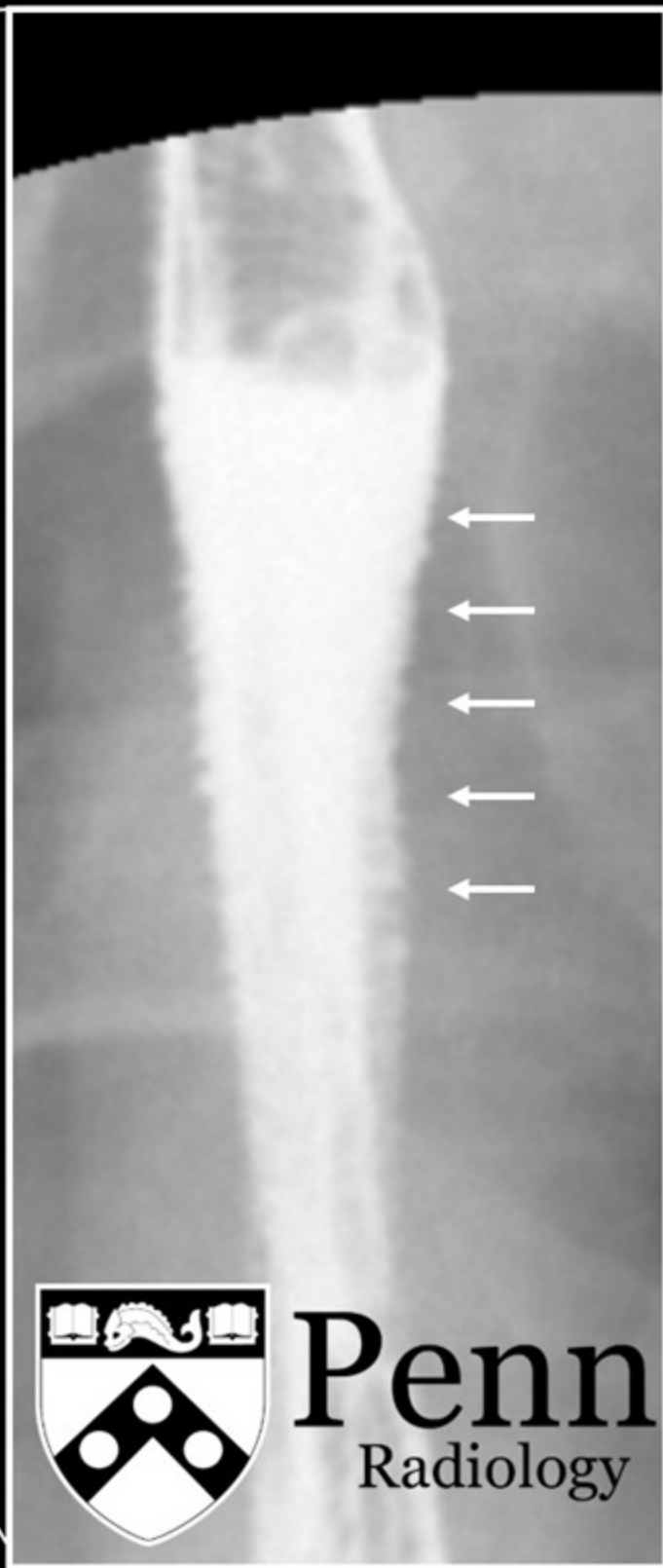
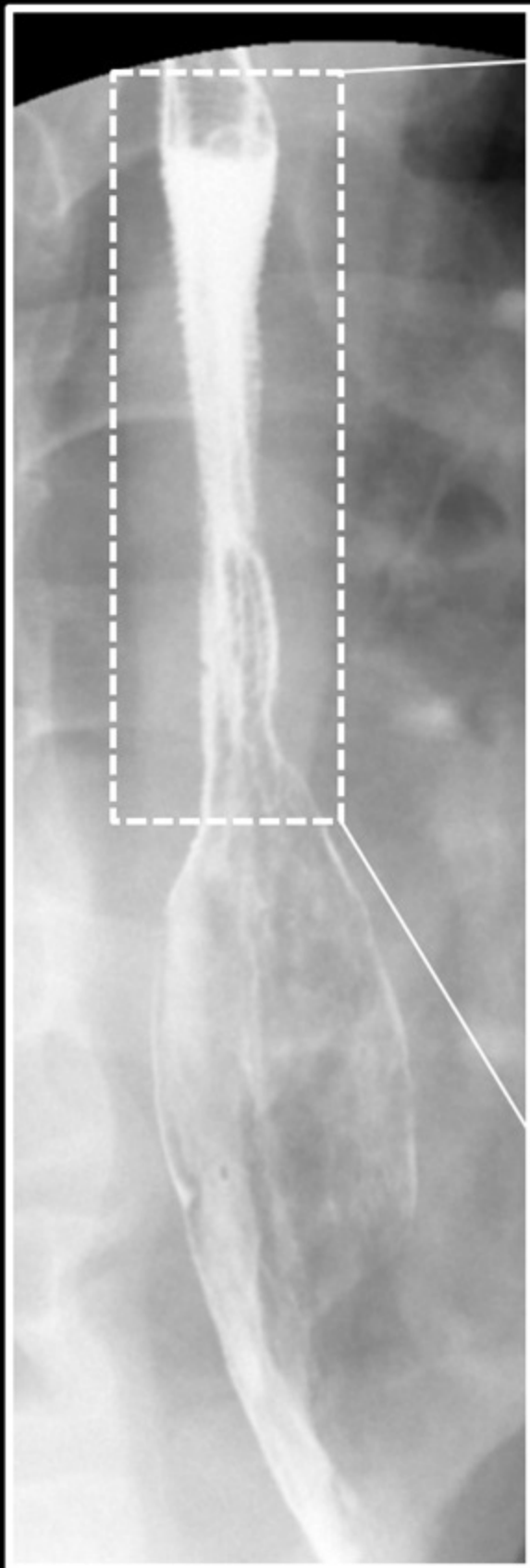
## History and esophagram images

Our appreciation is extended to Dr. Jamaal Benjamin, PhD, University of Pennsylvania Department of Radiology, for contributing this case.

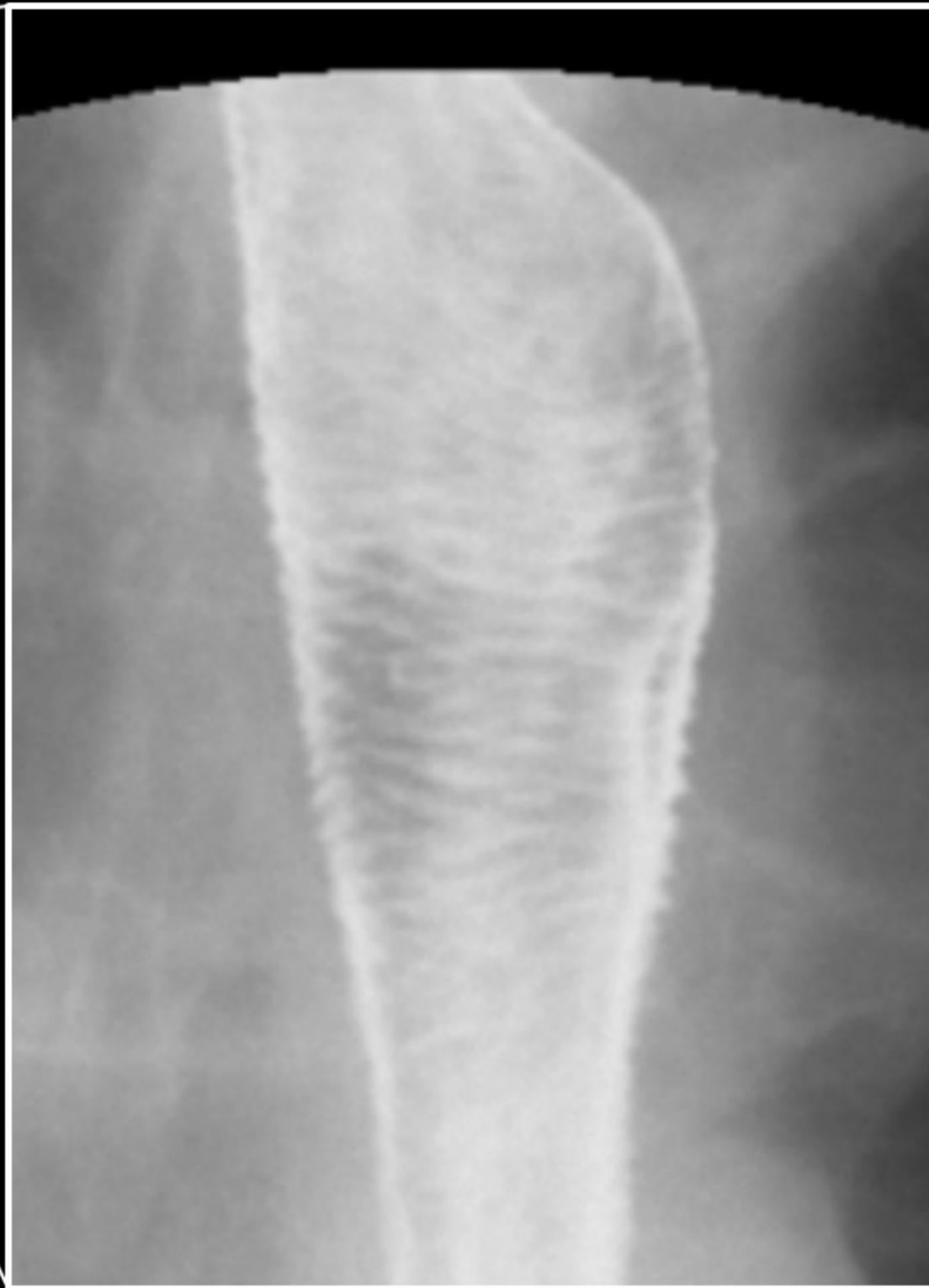
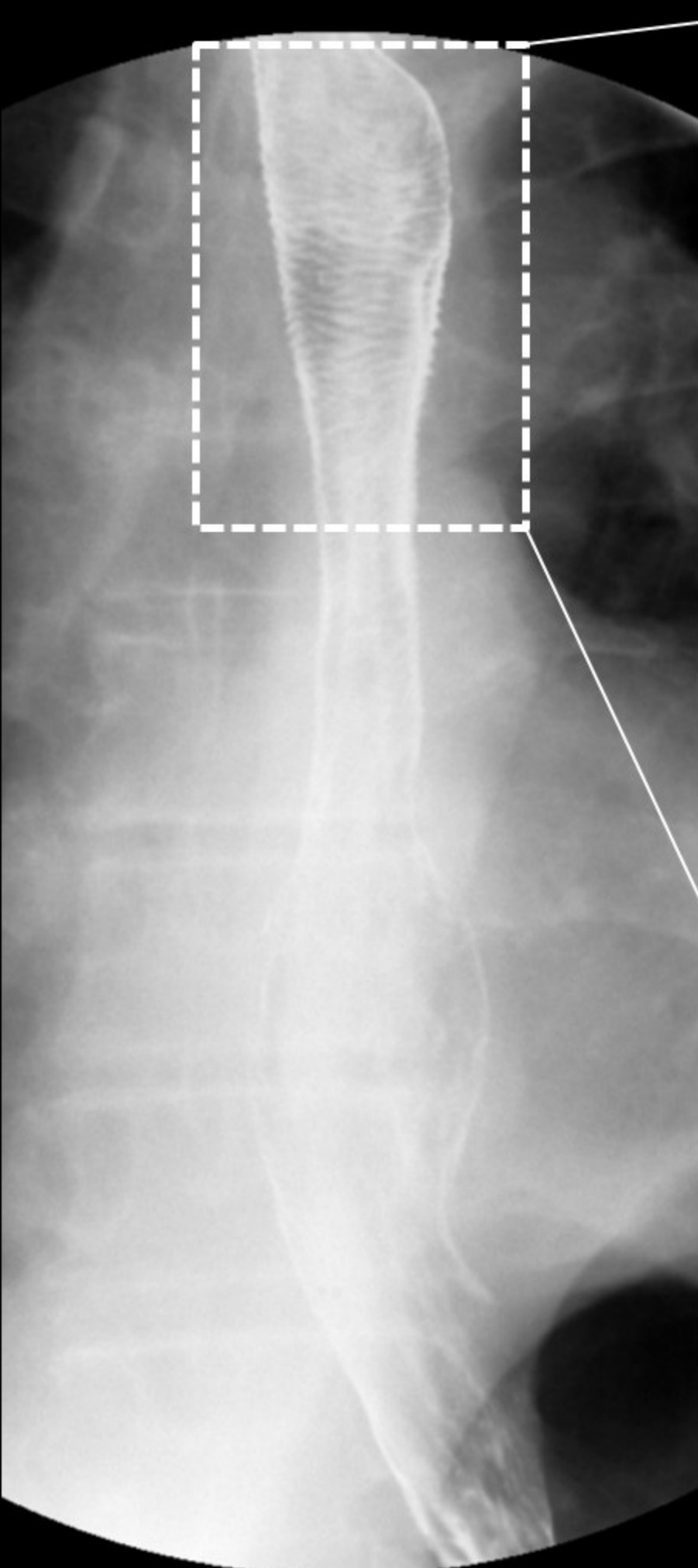
**History:** An 82-year-old man presents to his primary care provider with dysphagia to solids for the past eight months and approximately 20 lb of involuntary weight loss.

A double-contrast esophagram was ordered by the primary care provider. Erect left posterior oblique and prone right anterior oblique images of the esophagus, gastric cardia, and fundus are shown below. Click to enlarge.





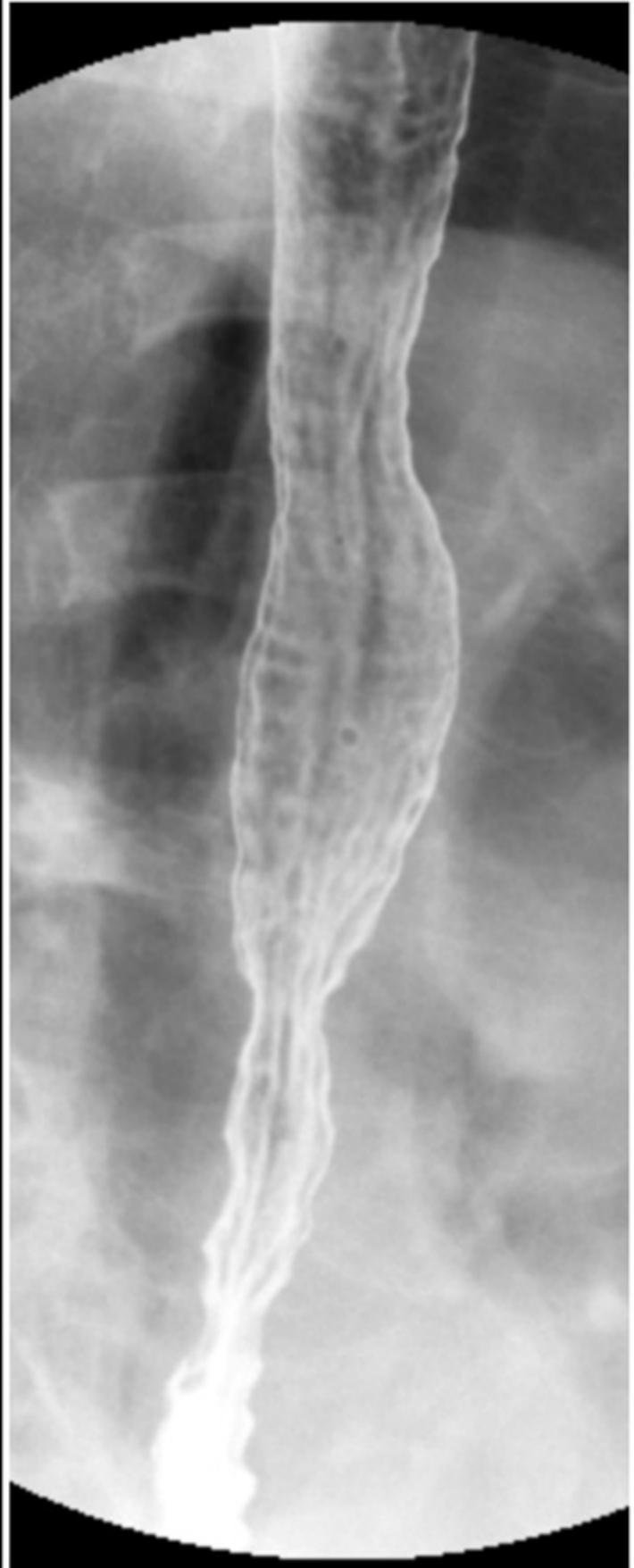
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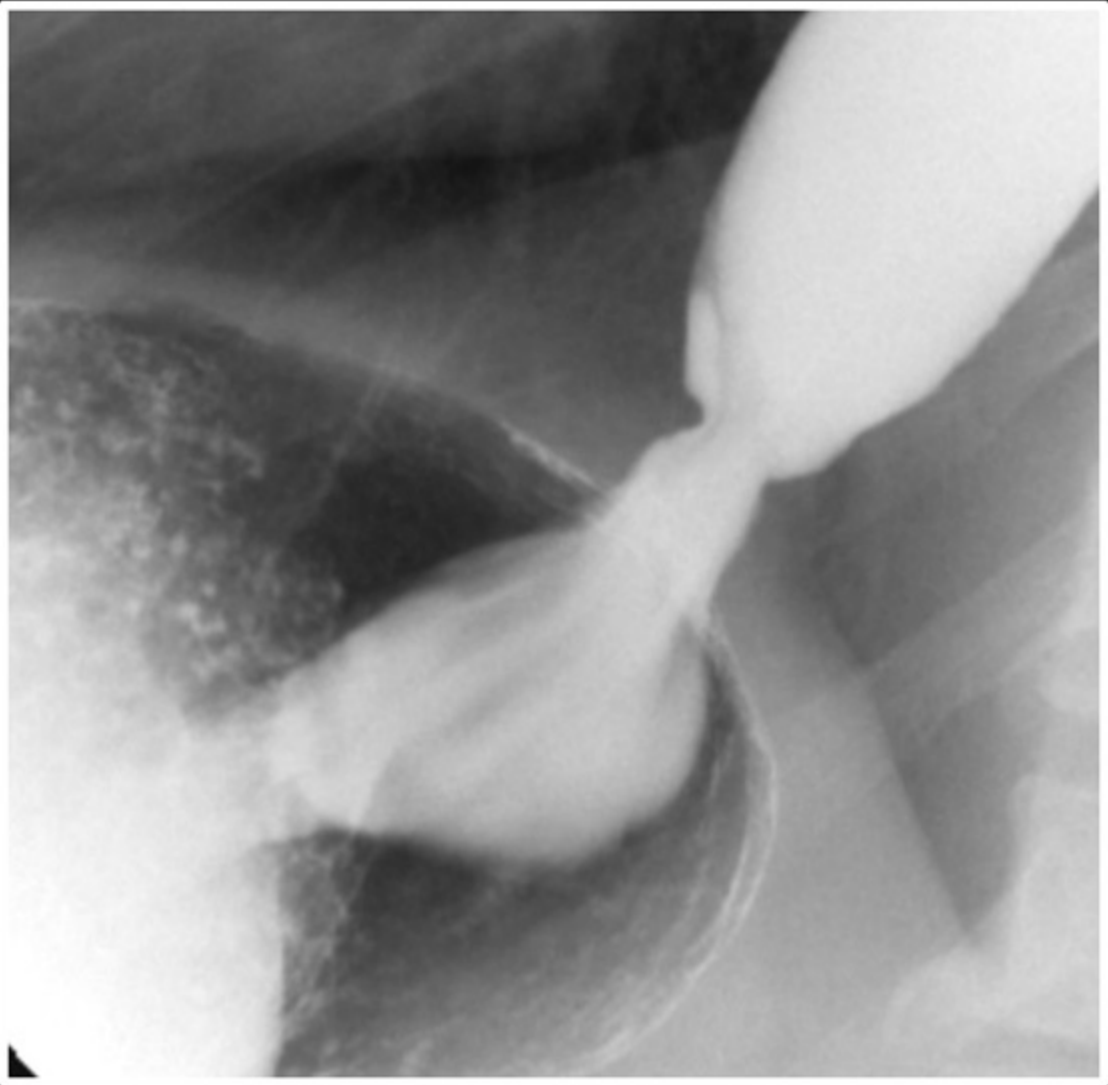
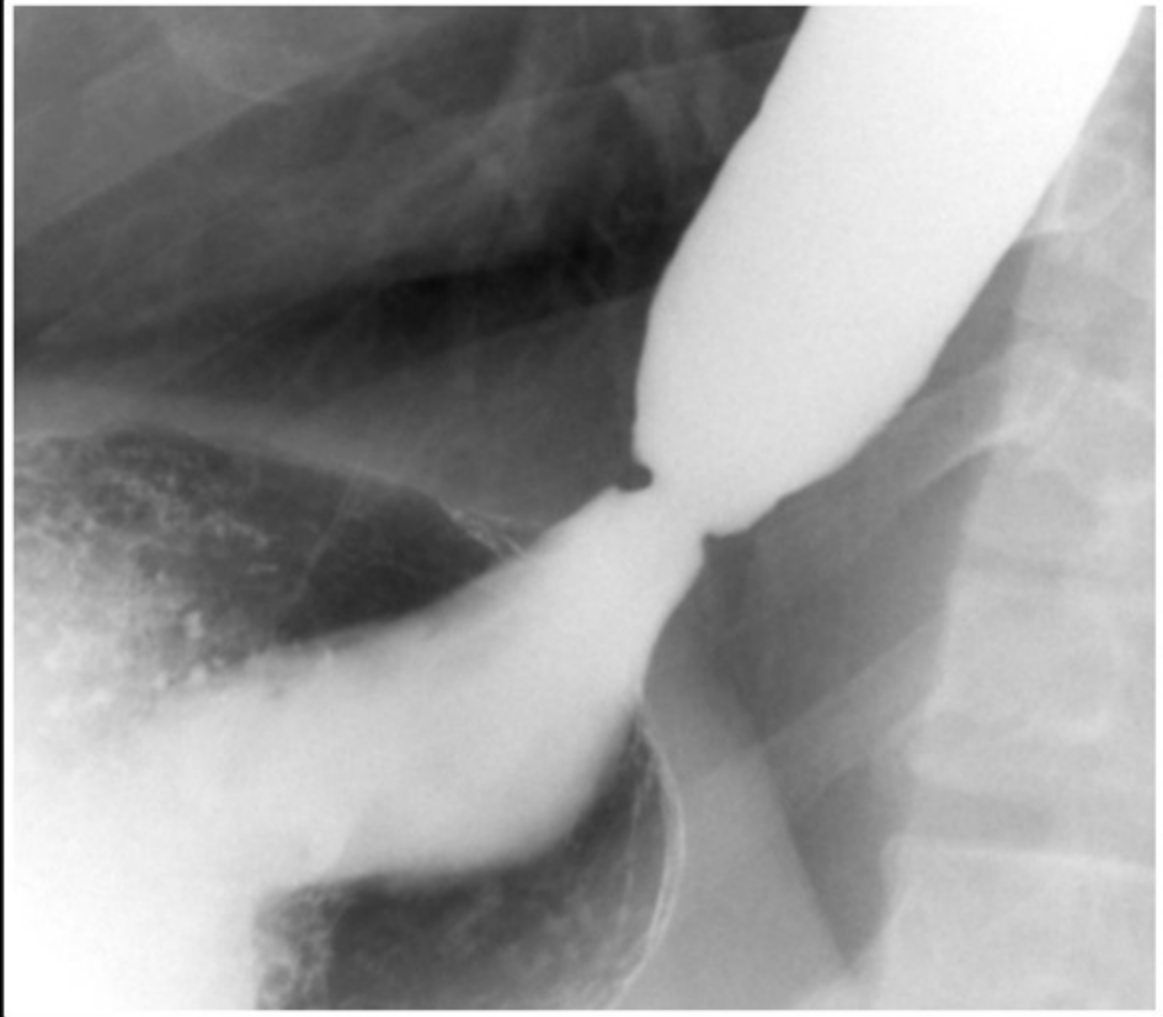
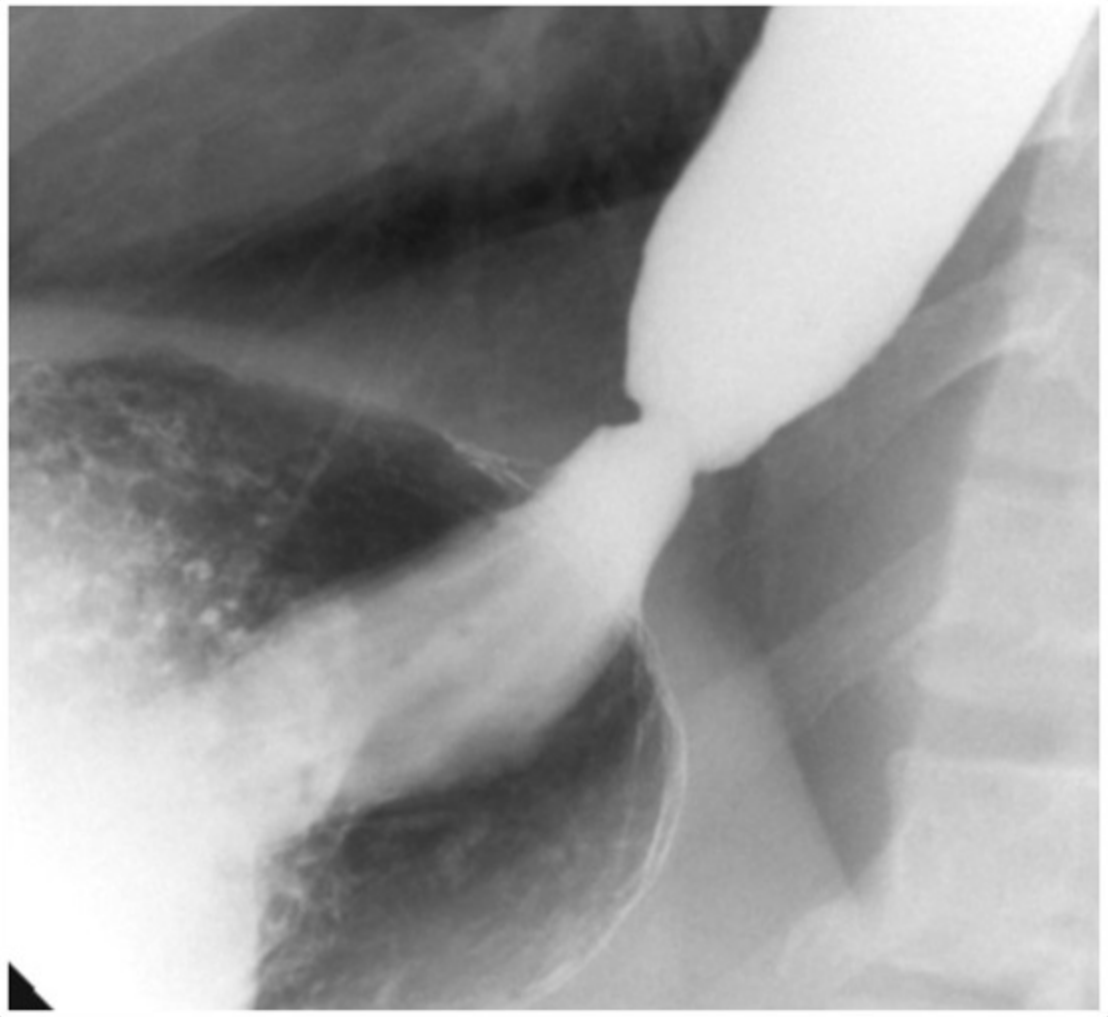


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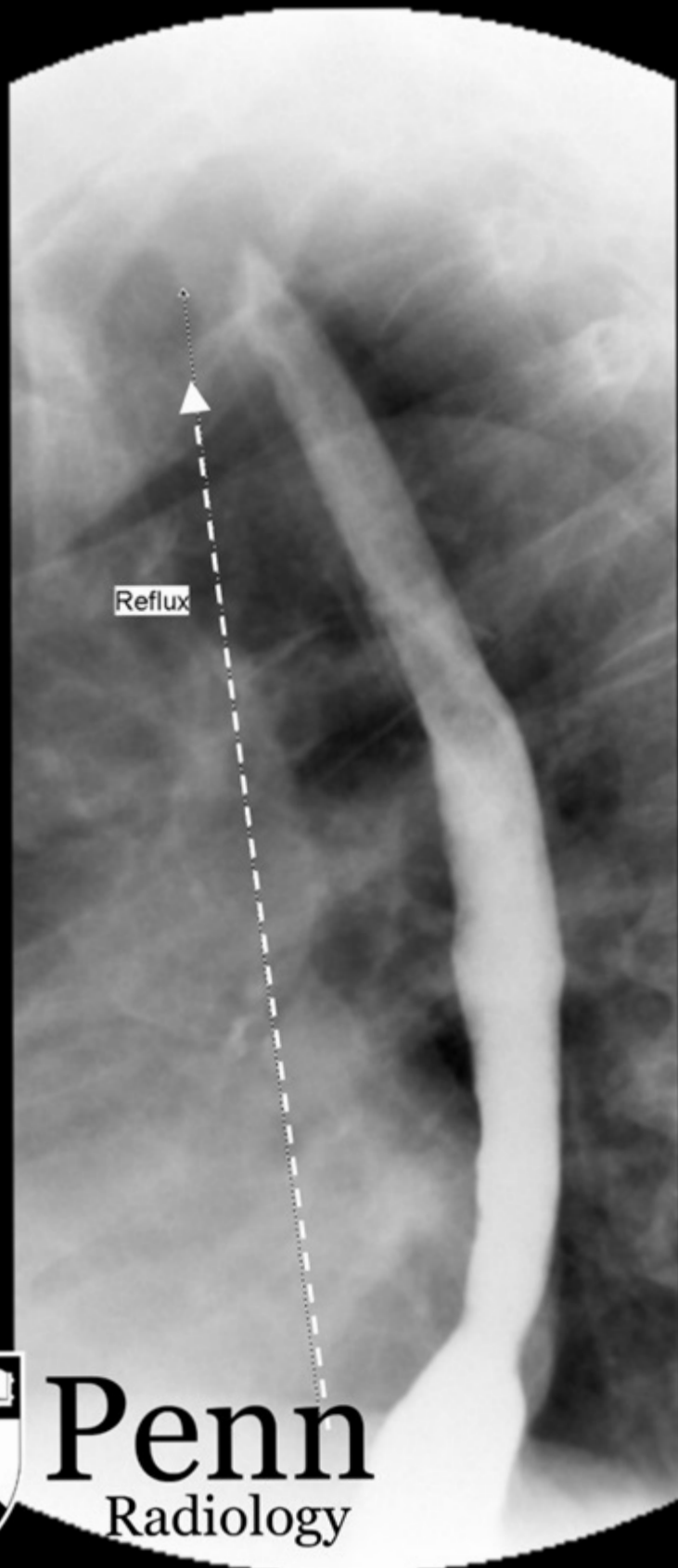


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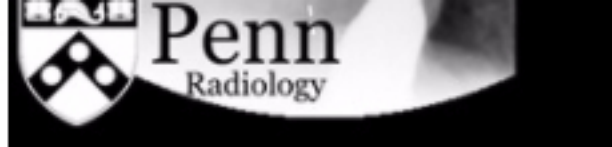
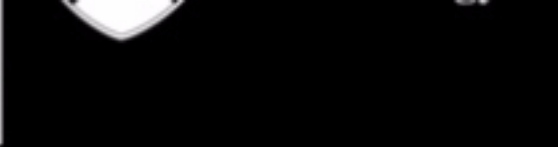


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Radiology





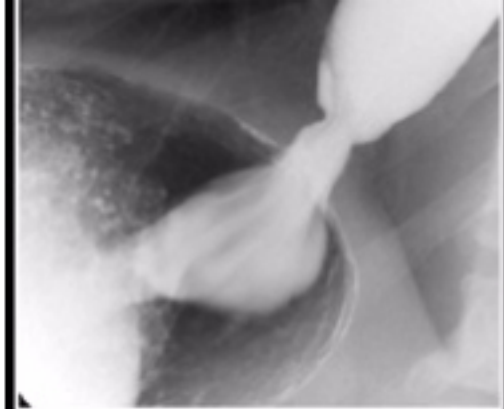
**There is a normal appearance to the mid/distal esophageal mucosa.**

☐ True

☐ False

The question above accounts for 16% of your total score for this case.

[VIEW YOUR SCORE](#)



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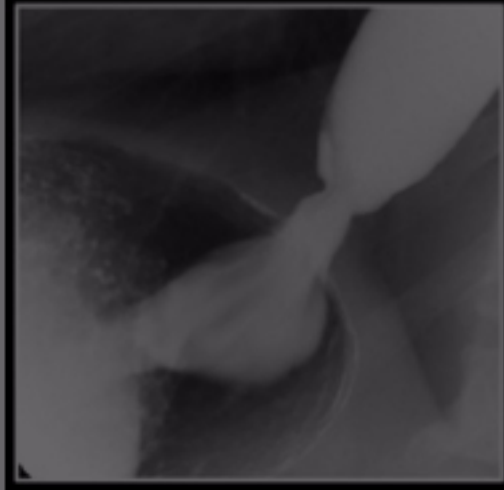
**There is a normal appearance to the mid/distal esophageal mucosa.**

☐ True

☒ False (correct!)

**[Explain this Answer]**

The question above accounts for 16% of your total score for this case.



There are longitudinally oriented folds in the esophagus. The normal esophagus should appear featureless on double-contrast esophagram.

There is a normal esophagus.

☐ True

☒ False (correct)

cosa.

**What is the best explanation for the longitudinally oriented folds in the esophagus (insert images and arrows).**

- ☐ Glycogenic acanthosis
- ☐ Spasm of muscularis mucosae
- ☐ Herpes esophagitis
- ☐ Esophageal varices

The question above accounts for 16% of your total score for this case.

What is the best explanation for the longitudinally oriented folds in the esophagus (insert images and arrows).



Horizontally

- ☐ Glycogenic acanthosis
- ☒ Spasm of muscularis mucosae (correct!)
- ☐ Herpes esophagitis
- ☐ Esophageal varices

[Explain this Answer]



Feline esophagus is secondary to spasm of the longitudinally muscle fibers.

## Additional questions

**How frequently is the so-called feline esophagus associated with gastroesophageal reflux disease?**

☐ 10% to 20%

☐ 30% to 50%

☐ 50% to 75%

☐ Nearly 100%

## Additional questions

How frequently is the so-called feline esophagus associated with gastroesophageal reflux disease?

☐ 10% to 20%

☐ 30% to 50%

☐ 50% to 75%

☒ Nearly 100% (correct!)

**The distal esophagus measures approximately 8 mm. In this patient with dysphagia, what is the most appropriate way to describe the focal narrowing of the esophagus?**

☐ Esophageal web

☐ Hiatal hernia

☐ Schatzki ring

☐ Esophageal varices

The distal esophagus measures approximately 8 mm. In this patient with dysphagia, what is the most appropriate way to describe the focal narrowing of the esophagus?

☐ Esophageal web

☐ Hiatal hernia

☒ Schatzki ring (correct!)

☐ Esophageal varices





X

The term Schatzki ring is reserved for symptomatic lower esophageal rings.

### [Explain this Answer]

The question above accounts for 17% of your total score for this case.

**What is the most common infectious cause of esophagitis?**

☐ *Cytomegalovirus* (CMV)

☐ *Candida albicans*

☐ Herpes simplex virus (HSV)

The question above accounts for 17% of your total score for this case.

**Where would a transition from squamous epithelium to columnar epithelium**

The question above accounts for 17% of your total score for this case.

## What is the most common infectious cause of esophagitis?

- ☐ *Cytomegalovirus* (CMV)
- ☒ *Candida albicans* (correct!)
- ☐ Herpes simplex virus (HSV)

### [Explain this Answer]

The question above accounts for 17% of your total score for this case.

## Where would a transition from squamous epithelium to columnar epithelium

The question above accounts for 17% of your total score for this case.

**Where would a transition from squamous epithelium to columnar epithelium normally be expected?**

☐ Mucosal A ring

☐ Mucosal B ring

☐ Mucosal C ring

☐ None of the above

**Where would a transition from squamous epithelium to columnar epithelium normally be expected?**

☐ Mucosal A ring

☒ Mucosal B ring (correct!)

☐ Mucosal C ring

☐ None of the above

**[Explain this Answer]**



## Findings

- There is nodularity of the distal esophagus, likely reflecting esophagitis. Additionally, several episodes of feline esophagus are noted -- a finding almost pathognomonic for gastroesophageal reflux disease.
- There is a small hiatal hernia with large volume gastroesophageal reflux to the level of the upper thoracic esophagus.
- A prominent mucosal ring is near the gastroesophageal junction, measuring approximately 8 mm in maximal luminal diameter. This finding could represent a Schatzki ring given the patient's clinical history.
- There is esophagitis of the distal esophagus.

## Differential diagnosis

- Feline esophagus

- Confirming given the patient's clinical history:
- There is esophagitis of the distal esophagus.

## Differential diagnosis

- Feline esophagus
- Focal spiculation -- normal variant in upper thoracic (upper third) esophagus
- Glycogenic acanthosis
- Reflux esophagitis scarring

**Diagnosis:** Feline esophagus

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# Feline esophagus

## Pathophysiology

- Feline esophagus is a **transient** phenomenon seen in the esophagus secondary to contraction of longitudinally oriented muscularis mucosae.
- The name is given because of the close resemblance to the transverse folds in cats.
- It is found almost exclusively in patients with gastroesophageal reflux disease (GERD).
- 80% of patients with feline esophagus will also have a hiatal hernia.
- Also known as the esophageal shiver.

## Epidemiology

- Feline esophagus is seen in approximately 2% to 9% of patients, although the transient nature of the finding may underestimate its prevalence.
- In a series from 2010, feline esophagus was detected in 9% of patients who underwent double-contrast esophagram.



- In a series from 2010, feline esophagus was detected in 9% of patients who underwent double-contrast esophagram.
- Of the patients with feline esophagus, transverse folds were seen in the middle and distal thirds 95% of the time.

## **Clinical presentation**

Patients with feline esophagus will typically present with GERD symptoms:

- Frequent heartburn
- Sour, bitter, or metallic taste in mouth
- Sore throat
- Coughing

Intermittent dysphagia is the most common presenting symptom for Schatzki's rings.

## **Imaging features**

## Double-contrast esophagram:

- Normal, distended esophagus has a smooth, featureless surface.
- Patients with feline esophagus will have transverse folds in the distal third of the esophagus.
- Folds are transient in nature and are approximately 1-2 mm in thickness, and they cross the width of the esophagus without interruption.
- Most commonly observed only during the reflux of barium (85%).

## Differential diagnoses

- Focal spiculation -- normal variant in upper thoracic (upper third) esophagus
- Glycogenic acanthosis
- Reflux esophagitis scarring

## Treatment



- Reflux esophagitis scarring

## Treatment

Treatment involves the management of GERD, which often utilizes the step-up approach:

- Mild and intermittent symptoms (fewer than two symptoms per week):
  - Lifestyle and dietary modifications, such as elevation of head of bed, weight loss, and avoiding alcohol and tobacco
  - Histamine 2 receptor antagonist (H2RAs)
- Severe symptoms:
  - Proton pump inhibitors if patient fails H2RAs
  - Management of underlying *Helicobacter pylori*
  - Surgery -- after failed maximal medical management: Nissen (360°), Toupet (270°), or Dor (180°) fundoplication

## References

**History:** A woman presents with flank pain.

Ultrasound images are shown below. Click to enlarge.





Lat/Mid Left Long

12  
Kidney

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P



Upper Left Long

12  
Kidney







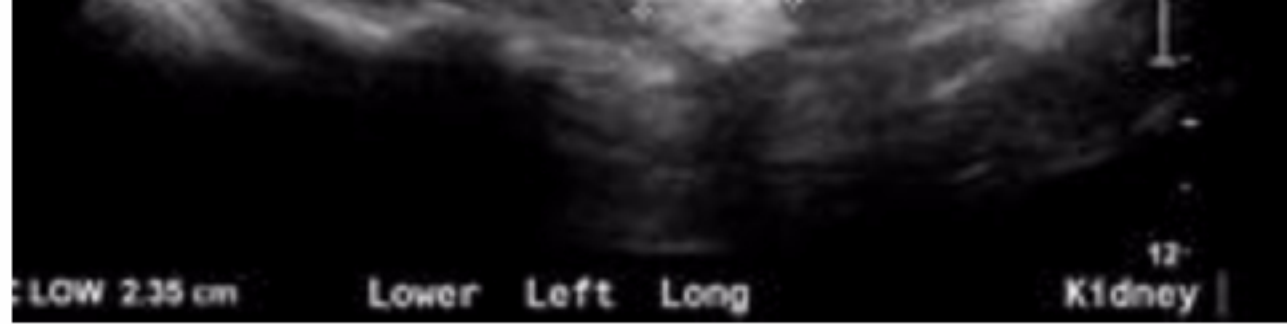
**There is hydronephrosis.**

☐ True

☐ False

The question above accounts for 16% of your total score for this case.

**A duplicated collecting system is seen.**



**There is hydronephrosis.**

☒ True (correct!)

☐ False

The question above accounts for 16% of your total score for this case.

**A duplicated collecting system is seen.**

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☐ True

☐ False

The question above accounts for 16% of your total score for this case.

**Collecting system gas is present.**

☐ True

☐ False

**A duplicated collecting system is seen.**

☐ True

☒ False (correct!)

The question above accounts for 16% of your total score for this case.

**Collecting system gas is present.**

☐ True

☒ False (correct!)

The question above accounts for 17% of your total score for this case.

**There is staghorn calculus.**

☐ True

☐ False

The question above accounts for 17% of your total score for this case.

[VIEW YOUR SCORE](#)

1

2

3

...

5

[next »](#)



The question above accounts for 17% of your total score for this case.

**There is staghorn calculus.**

☒ True (correct!)

☐ False

**[Explain this Answer]**

The question above accounts for 17% of your total score for this case.

[VIEW YOUR SCORE](#)

Densely shadowing material is seen in dilated calyces and also the renal pelvis, compatible with staghorn calculus.

☒ True (correct!)

☐ False

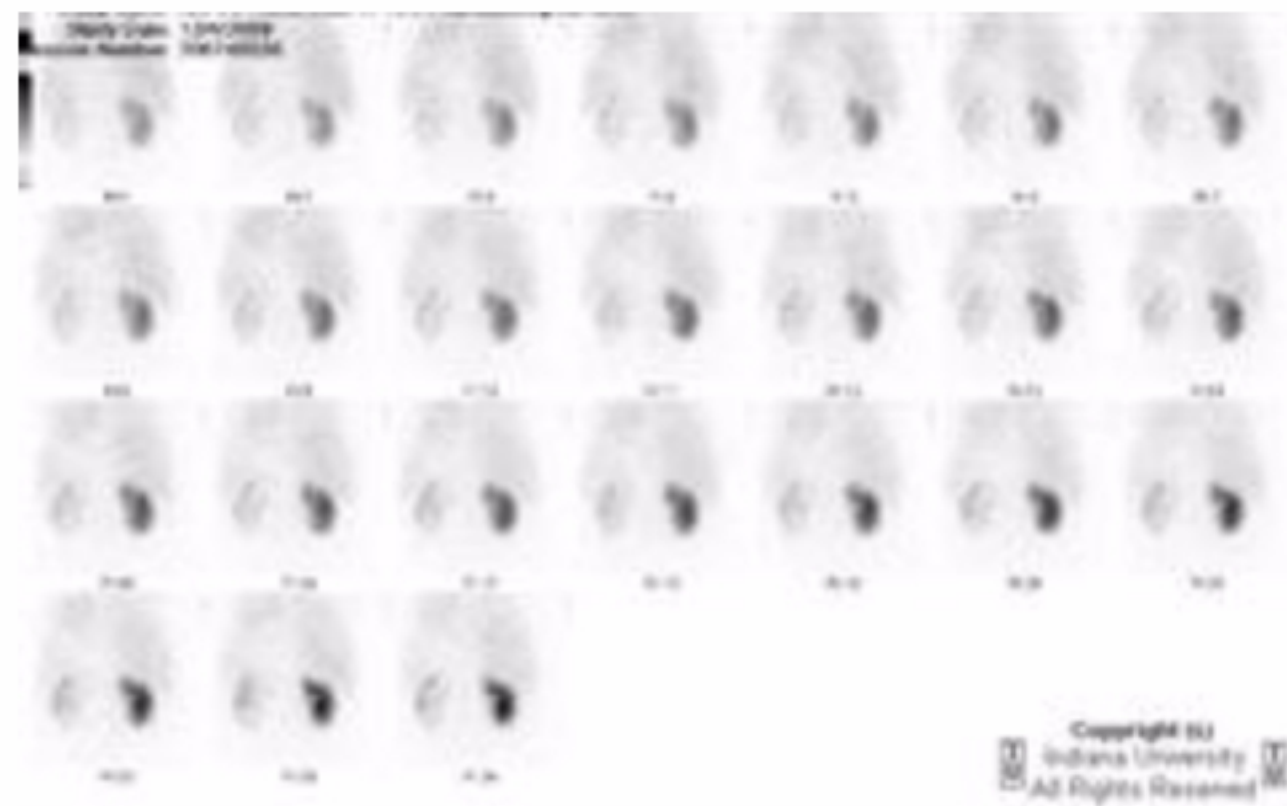
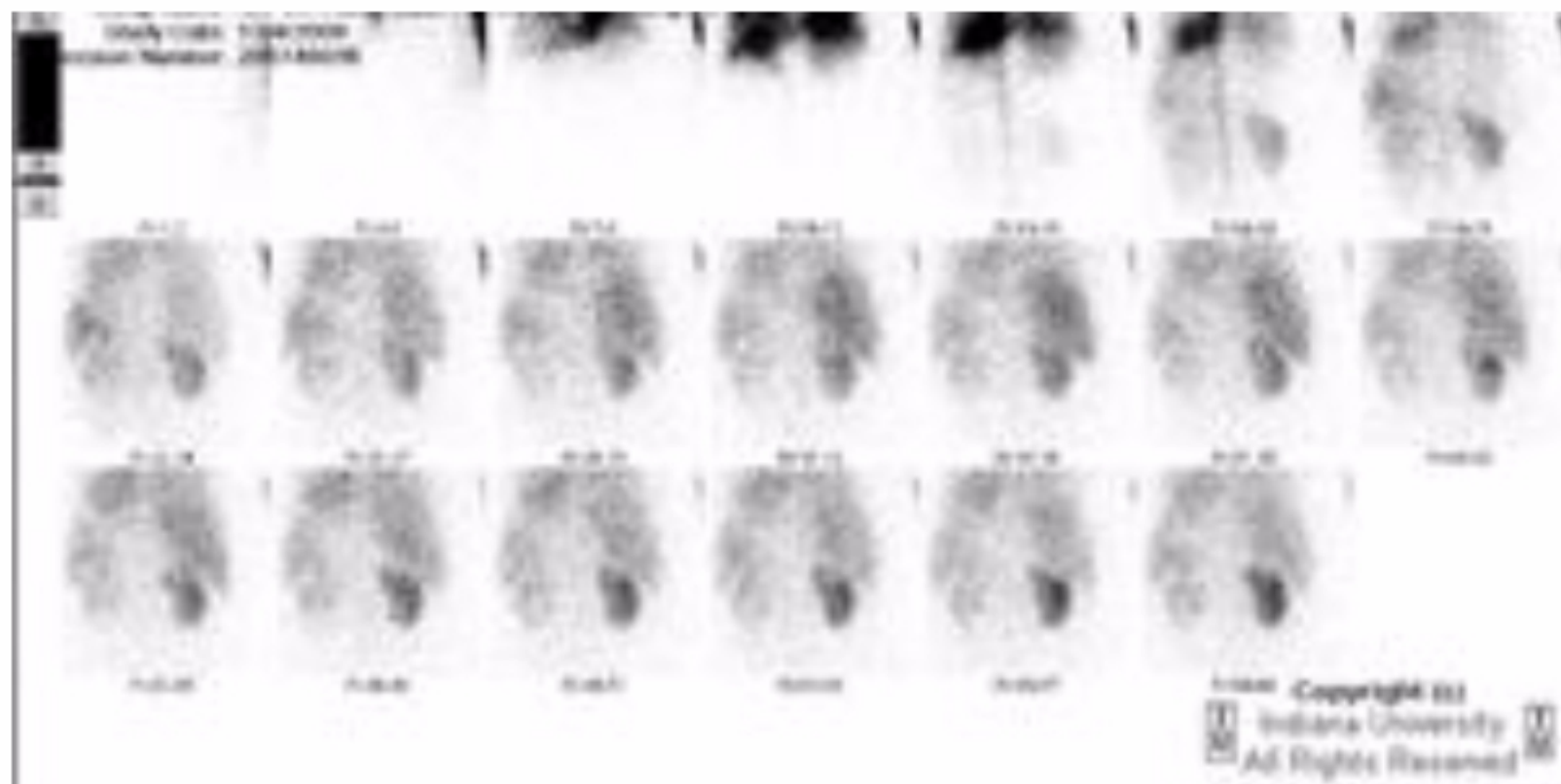
[**Explain this Answer**

The question above



## Nuclear medicine images

A nuclear medicine renal flow and function study also was performed. Images are displayed as if patient were prone. Click images to enlarge.



Fr1-3

Fr4-6

Fr7-9

Fr10-12

Fr13-15

Fr16-18

Fr19-21

Fr22-24

Fr25-27

Fr28-30

Fr31-33

Fr34-36

Fr37-39

Fr40-42

Fr43-45

Fr46-48

Fr49-51

Fr52-54

Fr55-57

Fr58-60

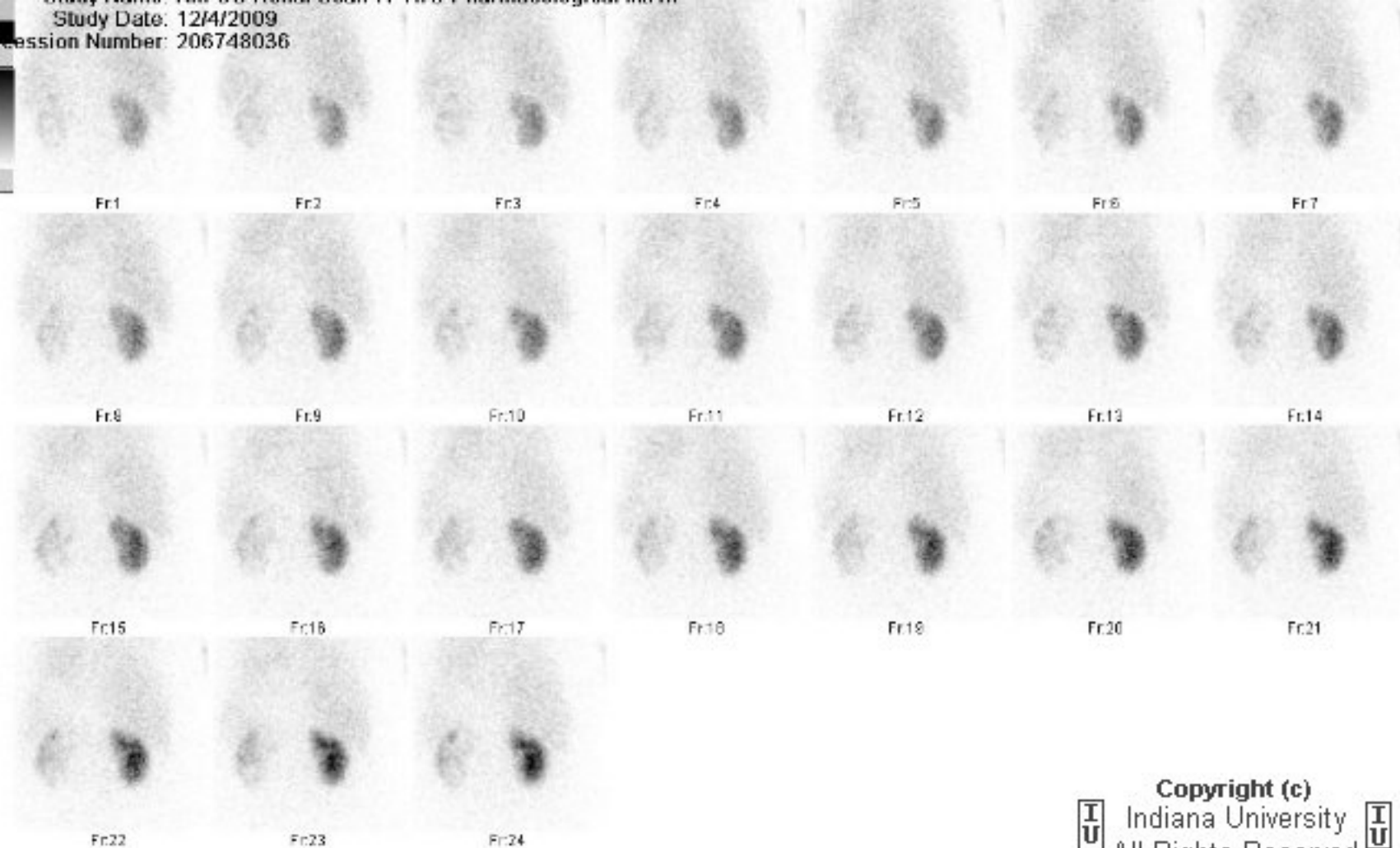
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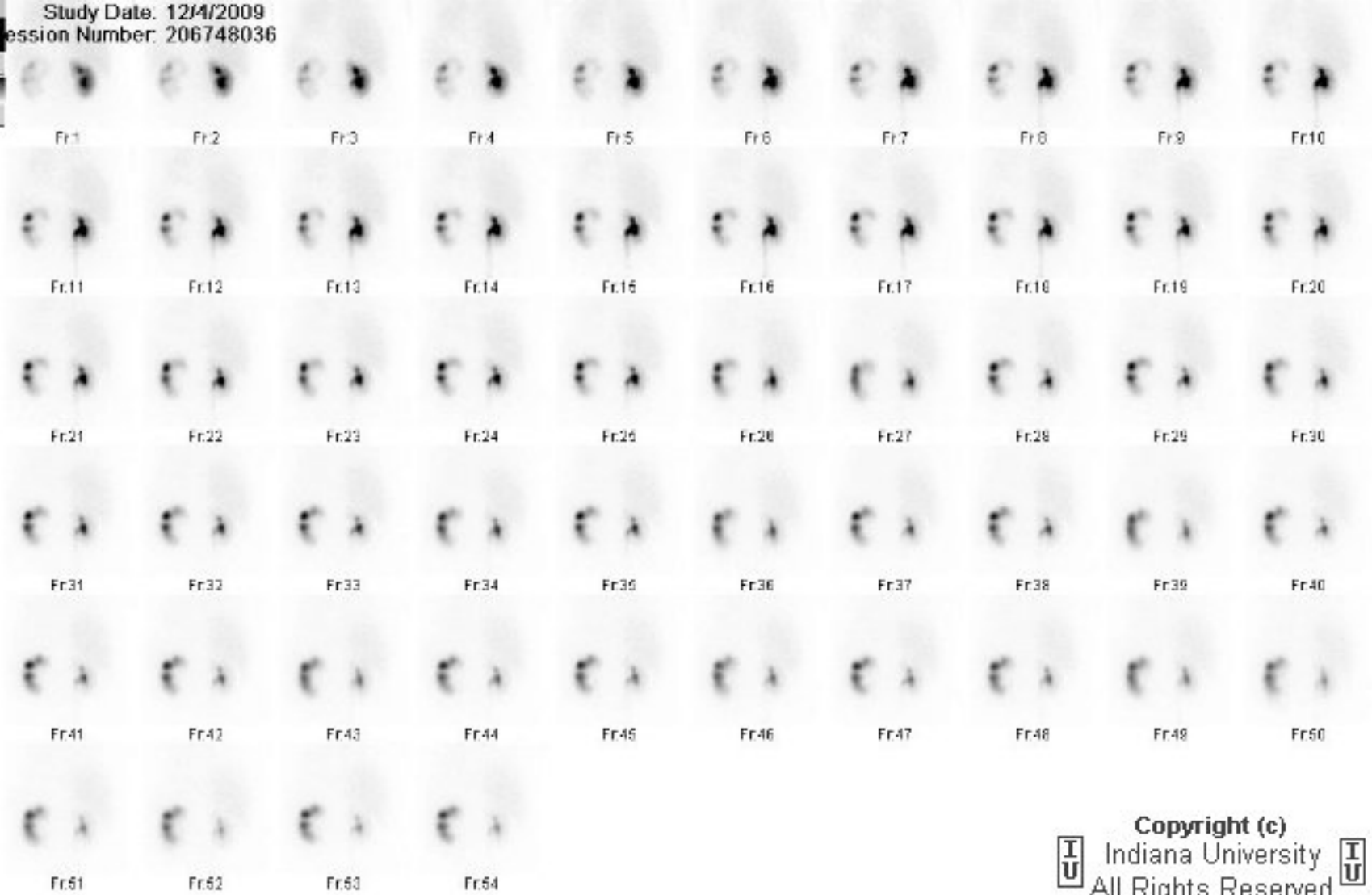


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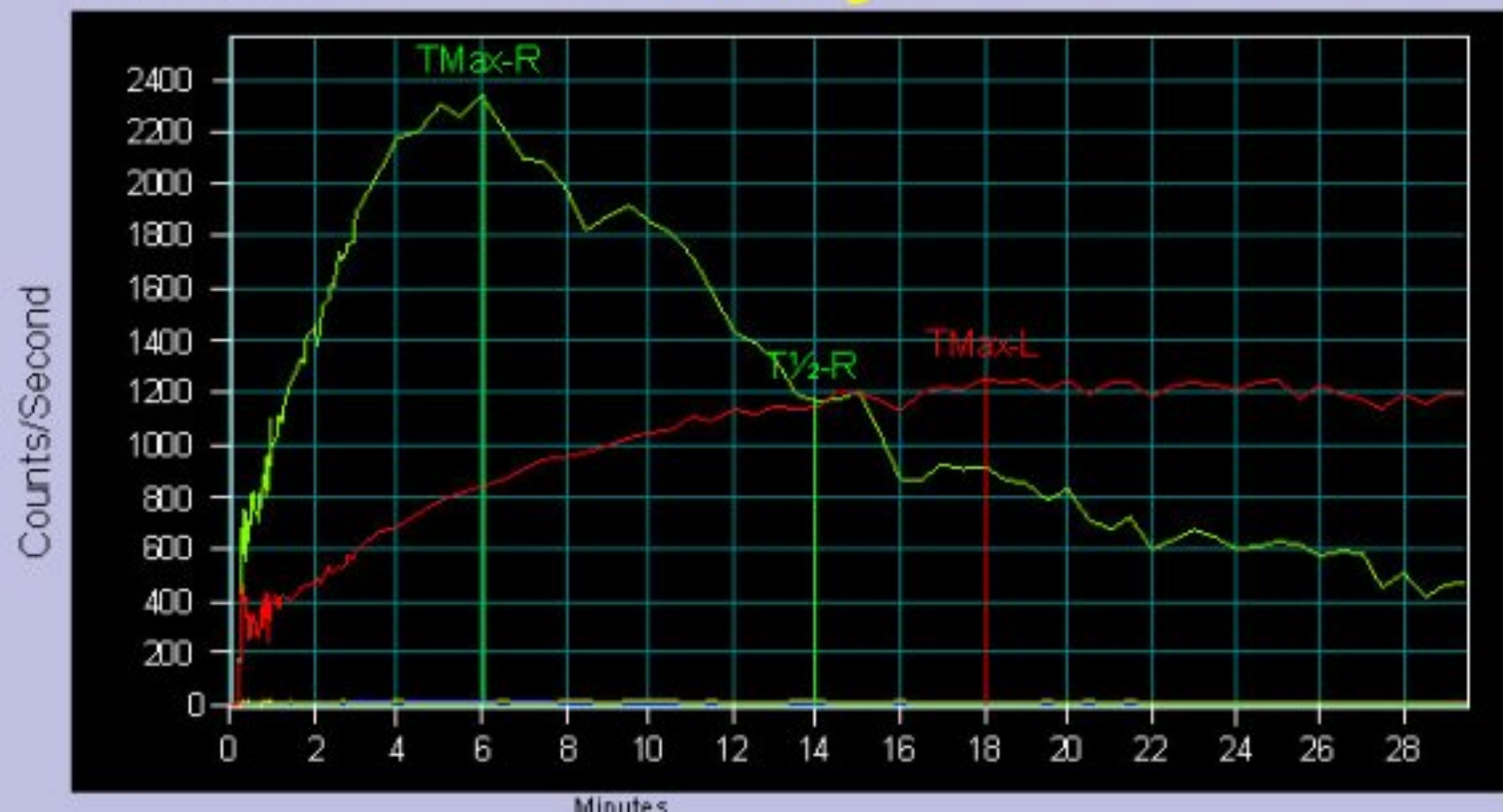








## Kidney



**There is delayed left renal perfusion.**

☐ True

☐ False

The question above accounts for 17% of your total score for this case.

**Findings are compatible with severe obstruction of the left renal collecting system.**

☐ True

☐ False

**There is delayed left renal perfusion.**

☒ True (correct!)

☐ False

The question above accounts for 17% of your total score for this case.

**Findings are compatible with severe obstruction of the left renal collecting system.**

☒ True (correct!)

☐ False





X

There is decreased cortical activity and central renal photopenic defect.

## Findings

There is moderate-to-severe hydronephrosis seen on renal ultrasound and large left staghorn calculus, best seen on scout radiograph. Nuclear medicine renal flow and function study shows delayed left renal perfusion with decreased cortical activity and central renal photopenic defect, suggestive of a dilated collecting system and severe obstruction of the left renal collecting system.

**Diagnosis:** Severe left renal collecting system obstruction with hydronephrosis and staghorn calculus

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## **Obstructive uropathy**

Obstructive uropathy is the blockage of urinary outflow at any level of the renal collecting system, from the kidney to the urethra, caused by congenital or acquired abnormalities and produces hydronephrosis or acute/chronic renal insufficiency.

- Congenital: Ureteropelvic junction (UPJ) obstruction, neonatal hydronephrosis, duplicated collecting system with ectopic insertion of ureter
- Acquired: Renal/urethral calculus, sequela of infection/trauma/radiation, extrinsic or intrinsic compression by tumor, bladder outlet obstruction

## **Clinical history**

Presenting symptoms commonly include renal colic, impaired renal function, and urinary tract infections.



## Imaging

Best imaging modality is radionuclide renal imaging (technetium-99m mercaptoacetyltriglycine [Tc-99m MAG3] renography).

- Other imaging modalities: CT (noncontrast and contrast-enhanced), ultrasound, intravenous pyelography (IVP)
- Best imaging for acute obstruction: IVP or noncontrast CT

### Imaging findings:

- High-grade obstruction with radionuclide renal flow and function studies (Tc-99m MAG3 renography):
  - Results can be affected by patient dehydration, renal insufficiency, decreased response to Lasix (longstanding use of Lasix), severely dilated nonobstructed collecting system, and immature kidneys.

- High-grade obstruction with radionuclide renal flow and function studies (Tc-99m MAG3 renography):
  - Results can be affected by patient dehydration, renal insufficiency, decreased response to Lasix (longstanding use of Lasix), severely dilated nonobstructed collecting system, and immature kidneys.
  - Diuretic renography is helpful to differentiate obstructed versus dilated, nonobstructed collected system.
  - Long-standing high-grade obstruction has no renal perfusion or function with flat renogram.
  - There is decreased or no washout from dilated collecting system (despite Lasix).
  - Renogram curve: Delayed time to cortical peak and cortical clearance with rising excretion slope.
  - Patients will have a variable degree of hydronephrosis.
  - There is reduced renal perfusion and function.
  - O'Reilly classification of obstructive uropathy (based on renal and collecting system washout curves):



collecting system washout curves):

<b>O'Reilly classification of obstructive uropathy</b>			
<b>Degree of obstruction</b>	<b>Curve type</b>	<b>Characteristics</b>	<b>Washout T <math>\frac{1}{2}</math></b>
<b>Normal</b>	I	Spontaneous washout of activity from collecting system	< 10 min
<b>High-grade obstruction</b>	II	Progressive rise in renal collecting system activity (despite Lasix)	> 20 min
<b>Dilated, nonobstructed</b>	IIIa	Activity rises until Lasix, then normal washout	< 10 min
<b>Partial obstruction</b>	IIIb	Activity rises until Lasix, washout ensues but slower than normal	
<b>Low-grade partial</b>		Questionable clinical significance	10-15 min
<b>Partial obstruction</b>		Clinically significant	15-20 min

- CT: CT shows collecting system dilation proximal to level of obstruction; secondary signs of dysfunction (perinephric stranding, etc.) correspond to severity with or without treatment.
  - Noncontrast CT: Identifies calculi; has low sensitivity (27% to 42%) for noncalcareous hydronephrosis.
  - Contrast-enhanced CT: Can show tumors or other causes of extrinsic compression.
  - Intravenous pyelogram (IVP): Delayed nephrogram and collecting system opacification, dilated collecting system, and dilated ureter with column of contrast to level of ureteral obstruction (if ureteral obstruction is present).
- MRI: Helps to evaluate dilated upper urinary tract without radiation exposure.
  - T2-weighted imaging: Shows anatomy (dilated, fluid-filled urinary system) without contrast administration and regardless of renal function.
  - Dynamic function MR urography: Quantifies renal parenchymal function even if poor renal function or gross hydronephrosis.



- poor renal function or gross hydronephrosis.
- Renal ultrasound: Shows hydronephrosis with or without hyperechoic, shadowing stone. Diuretic-enhanced Doppler will show elevated resistive index and pulsatile index.

## Differential diagnoses

Imaging findings of other common differential diagnoses:

- Dilated, nonobstructed collecting system: Hydronephrosis minimized with postural drainage maneuver
- Renal vein thrombosis: Decreased renal blood flow/function, rising nephrogram, decreased visualization of collecting system
- Renal artery thrombosis: Small kidney; delayed nephrogram; decreased excretion, especially with administration of angiotensin converting enzyme inhibitors (ACE) inhibitor
- Acute tubular necrosis: Blood flow normal/mildly decreased, rising nephrogram
- Vesicoureteral reflux: Present with or without obstruction, catheterize bladder during

especially with administration of angiotensin converting enzyme inhibitors (ACE) inhibitor

- Acute tubular necrosis: Blood flow normal/mildly decreased, rising nephrogram
- Vesicoureteral reflux: Present with or without obstruction, catheterize bladder during lasix renography to avoid mimic of obstruction
- Medical renal disease: Impaired renal function and blood flow

## **Treatment and management of high-grade obstruction**

- Prognosis: Can cause impaired renal function and infections.
- Treatment involves mechanical relief of obstruction. Options include stent placement, nephrostomy, pyeloplasty, urinary diversion, and lithotripsy.

## **References**

1. Boubaker A, Prior JO, Meuwly JY, Bischof-Delaloye A. Radionuclide investigations of the urinary tract in the era of multimodality imaging. J Nucl Med. 2006;47(11):1819-

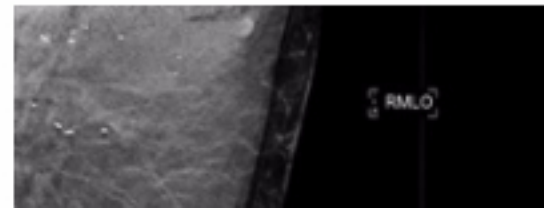


## History and mammography images

Our appreciation is extended to Dr. Erica Alexander, University of Pennsylvania Department of Radiology, for contributing this case.

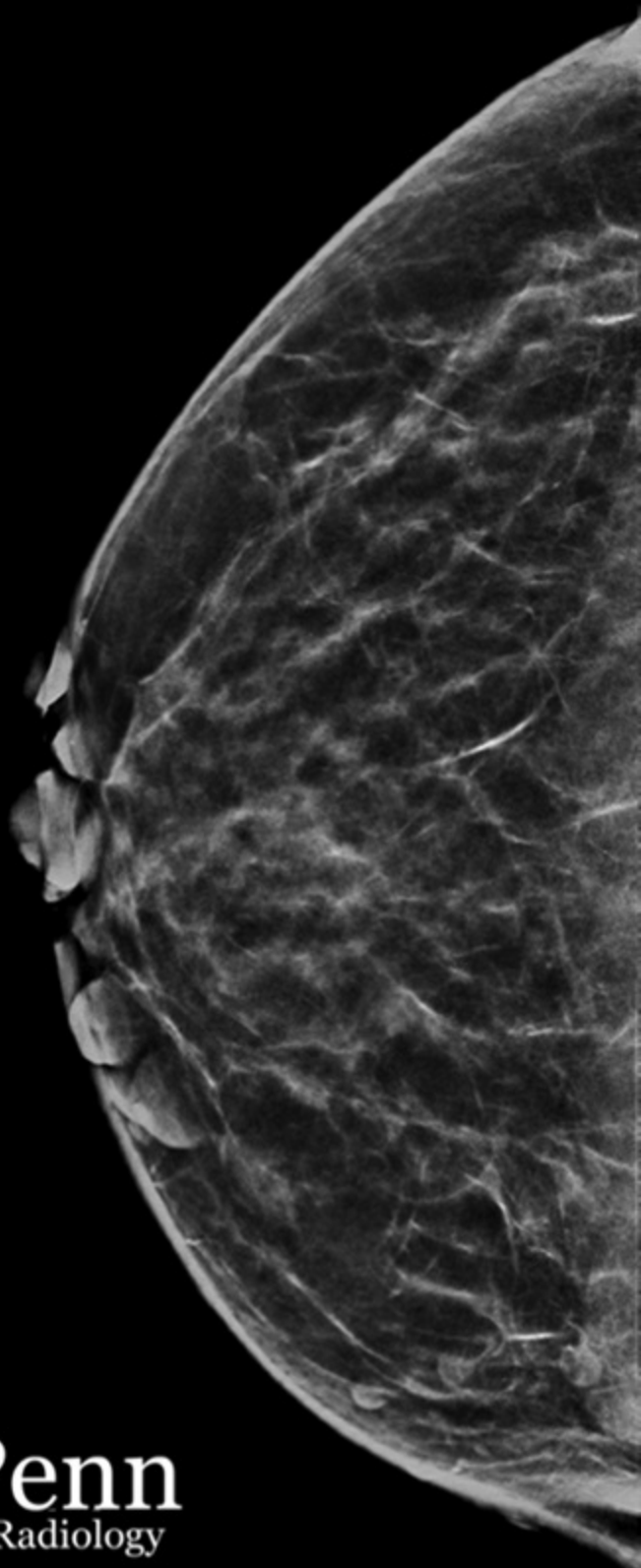
**History:** A 40-year-old asymptomatic woman presents for screening mammography. Additional history temporarily withheld.

Bilateral mammography images are shown below. Click to enlarge. In order: left and right craniocaudal (CC) and mediolateral oblique (MLO) images.



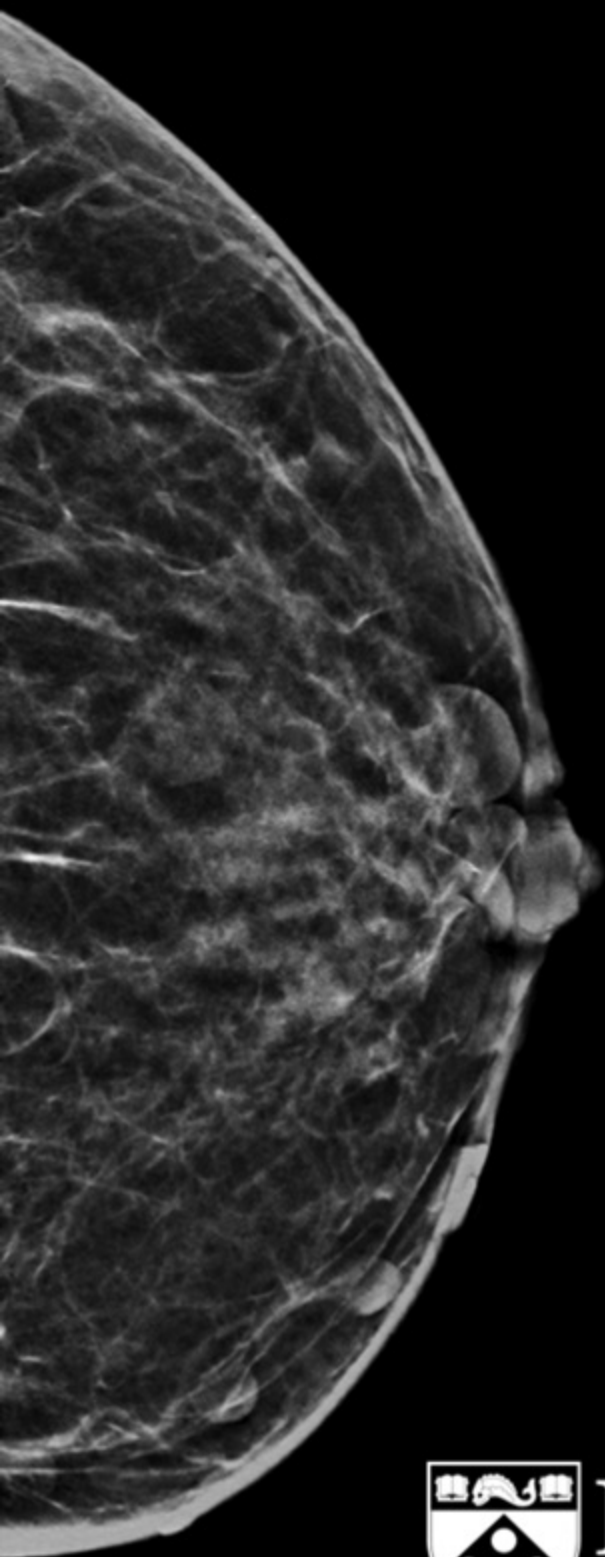


TT  
LK LCC



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RCC



Penn  
Radiology

LMLO



Penn  
Radiology

RMLO



Penn  
Radiology

**What BI-RADs classification for breast density is most appropriate for this patient?**

- ☐ Predominantly fat
- ☐ Scattered fibroglandular densities
- ☐ Heterogeneously dense
- ☐ Extremely dense



**What BI-RADs classification for breast density is most appropriate for this patient?**

- ☐ Predominantly fat
- ☐ Scattered fibroglandular densities
- ☐ Heterogeneously dense
- ☒ Extremely dense (correct!)

The question above accounts for 20% of your total score for this case.

The question above accounts for 20% of your total score for this case.

**In what portion of the breasts are the findings most likely located?**

☐ Skin

☐ Fatty tissue

☐ Nipple

☐ Lobules

The question above accounts for 20% of your total score for this case.

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**In what portion of the breasts are the findings most likely located?**

☒ Skin (correct!)

☐ Fatty tissue

☐ Nipple

☐ Lobules

The question above accounts for 20% of your total score for this case.

The question above accounts for 20% of your total score for this case.

**What is the most appropriate next step to work up these findings?**

☐ Ultrasound

☐ Mammogram

☐ Biopsy

☐ No further workup is needed

The question above accounts for 20% of your total score for this case.

**What is the most appropriate next step to work up these findings?**

☐ Ultrasound

☐ Mammogram

☐ Biopsy

☒ No further workup is needed (correct!)



The question above accounts for 20% of your total score for this case.

**What is the most likely diagnosis?**

- ☐ Steatocystoma multiplex
- ☐ Neurofibromatosis type 1
- ☐ Scleroderma
- ☐ Pseudoxanthoma elasticum

The question above accounts for 15% of your total score for this case.

The question above accounts for 20% of your total score for this case.

### What is the most likely diagnosis?

- ☐ Steatocystoma multiplex
- ☒ Neurofibromatosis type 1 (correct!)
- ☐ Scleroderma
- ☐ Pseudoxanthoma elasticum

The question above accounts for 15% of your total score for this case.

## Additional history and questions

Additional information: The patient has a history of neurofibromatosis.

**The multiplicity of these lesions makes them more likely to be benign.**

☐ True

☐ False

## Additional history and questions

Additional information: The patient has a history of neurofibromatosis.

**The multiplicity of these lesions makes them more likely to be benign.**

☒ True (correct!)

☐ False

## Additional history and questions

Additional information



The multiplicity

☒ True (correct)

☐ False

[Explain this Answer]

The question above

benign.



**[Explain this Answer]**

The question above accounts for 10% of your total score for this case.

**This disorder is associated with a higher rate of breast malignancies.**

☐ True

☐ False

The question above accounts for 15% of your total score for this case.

[VIEW YOUR SCORE](#)

The question above accounts for 10% of your total score for this case.

**This disorder is associated with a higher rate of breast malignancies.**

☒ True (correct!)

☐ False

**[Explain this Answer]**

The question above accounts for 15% of your total score for this case.

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Sharif and colleagues retrospectively evaluated a cohort of 304 patients between 40 and 49 years of age with neurofibromatosis type 1. The study sample had a 5.8% risk of developing breast cancer, compared with a 1.5% risk for the general population.

☐ False

The question above

**This disorder i**

☒ True (correct!

☐ False

**[Explain this Answ**

The question above

**ancies.**

## Findings

The breasts are extremely dense. No significant masses, calcifications, or other abnormal findings are seen. There are multiple, round, cutaneous, bilateral masses, all similar in appearance, in keeping with the patient's history of neurofibromatosis.

## Differential diagnosis

- Multiple breast cysts
- Multiple fibroadenomas
- Neurofibromatosis
- Bilateral multifocal cancer

**Diagnosis:** Neurofibromatosis; overall BI-RADS category: 1 -- negative

# Neurofibromatosis

## Pathophysiology

Neurofibromatosis type 1 (NF-1) is an autosomal dominant genetic disorder that is caused by a mutation in the NF-1 tumor suppressor gene. The disease can manifest as multiple rounded neurofibromas of various sizes on the skin of the breast and the nipple-areolar complex.

## Epidemiology

The incidence of neurofibromatosis is 1 in 2,500 to 3,000. Approximately half of the cases are inherited as an autosomal dominant condition; the other 50% of cases are due to a new mutation.

## Clinical presentation



## Clinical presentation

The diagnosis of neurofibromatosis type 1 requires two or more of the following criteria:

- More than six café au lait spots
- Two or more neurofibromas or one plexiform neurofibroma
- Optic nerve glioma
- Distinctive osseous lesion
- Sphenoid wing dysplasia
- Two or more Lisch nodules
- Axillary or inguinal freckling
- A primary relative with NF-1

Other health/cognitive effects associated with NF-1 include learning disabilities (range, 30% to 60%) and an increased incident of neoplasms. Sharif and colleagues retrospectively

Other health/cognitive effects associated with NF-1 include learning disabilities (range, 30% to 60%) and an increased incident of neoplasms. Sharif and colleagues retrospectively evaluated a cohort of 304 patients between 40 and 49 years of age with NF-1. The patients had a 5.8% risk of developing breast cancer, compared with a 1.5% risk for the general population. The increased risk highlights the importance of breast cancer screening with mammography in this patient population.

## **Imaging findings**

- **Mammography:** Neurofibromas appear as well-defined benign-appearing cutaneous masses. They tend to be periareolar in location. The neurofibromas typically have a lucent halo, which is created by air surrounding the nodule being compressed against the surface of the breast during the mammography procedure.
- **Ultrasound:** Neurofibromas appear as well-defined hypoechoic masses with posterior acoustic enhancement. If they are in the subcutaneous tissue, the features are similar



lucent halo, which is created by air surrounding the nodule being compressed against the surface of the breast during the mammography procedure.

- Ultrasound: Neurofibromas appear as well-defined hypoechoic masses with posterior acoustic enhancement. If they are in the subcutaneous tissue, the features are similar to that of fibroadenomas.

## Treatment

There is no single treatment for NF-1. Patients are treated with a combination of supportive and surgical therapies for particular tumors, based on their location and associated health effects.

## References

1. Cao MM, Hoyt AC, Bassett LW. Mammographic signs of systemic disease. *Radiographics*. 2011;31(4):1085-1100.



Table 1

Proposed classification of focal hand lesions at our institution into benign, intermediate-grade and malignant lesions

Lesion	Margins	T1w signal	T2w signal	Crossing compartments/involvement of adjacent structures	Bony destruction	Enhancement pattern
Benign						
Ganglion cyst	Smooth	Homogeneously hypointense	Homogeneously hyperintense (fluid)	Nil	NA	Rim enhancement, no internal enhancement
Epidermal cyst	Smooth	Homogeneously hypointense	Homogeneous hyperintense (fluid)	Nil	NA	Rim enhancement, no internal enhancement
Fibroma of tendon sheath	Lobulated, preserved fat planes	Intermediate	Heterogeneously hyperintense with low signal areas	Nil	Nil	Small eccentric focus of enhancement
Focal nodular synovitis	Fairly smooth	Hypointense	Homogeneously hypointense	Nil	Nil	No significant enhancement
Nodular fasciitis	Lobulated	Intermediate	Heterogeneously hyperintense	Crosses compartments (intra-and extra-compartmental)	NA	Heterogeneous, central non-enhancing foci
Lipoma	Lobulated, insinuating margins, preserved fat planes	Hyperintense	Homogeneous fat signal intensity	Crosses compartments (intra-and extra-compartmental, involves thenar and mid-palmar spaces)	Nil	Enhancement of thin internal septae only
Lipofibromatous hamartoma	Lobulated	Hyperintense with curvilinear low signal structures within	Homogeneous fat signal intensity surrounding neurovascular component	Nil	Nil	Linear enhancement of neurovascular components within the lesion
Haemangioma	Lobulated, insinuating margins	Slightly hyperintense	Heterogeneously hyperintense	Crosses compartments (intra-and extra-compartmental)	Nil	Avid, near homogenous post contrast enhancement
Schwannoma	Smooth	Isointense (to muscle)	Heterogeneously hyperintense, target sign	Nil	NA	Avid, near homogenous
Intermediate-grade (histologically benign but locally aggressive with potential for recurrence)						
Neurofibroma	Lobulated	Isointense	Heterogeneous	Crosses compartments (intra-and extra-compartmental)	Pressure erosion	Avid, near homogeneous
Desmoplastic fibroblastoma	Lobulated, loss of fat planes	Hypointense	Heterogeneous low signal	Encases adjacent extensor tendon	Nil	Heterogeneous internal and capsule enhancement
GCTTS	Lobulated	Intermediate	Heterogeneous intermediate-high with low signal areas due to haemosiderin deposits	Crosses compartments (intra-and extra-compartmental)	Pressure erosion	Avid, near homogeneous
Glomus tumour	Smooth	Intermediate	Homogeneously hyperintense	Crosses compartments (intra-and extra-compartmental)	Pressure erosion	Avid, homogenous
Malignant						
Undifferentiated pleomorphic sarcoma	Irregular	Hypointense	Heterogeneously hyperintense	Crosses compartments (intra-and extra-compartmental, areas of infiltration into adjacent muscle)	Nil	Heterogeneous
SCC	Infiltrative	Hypointense/intermediate	Heterogeneously hyperintense	Invading adjacent muscle, skin, bone	Yes	Heterogeneous
Malignant bone lesions	Infiltrative	Hypooointense/intermediate	Heterogeneously hyperintense	Invading adjacent muscle, skin, bone, soft tissue component	Yes	Heterogeneous
Fibrosarcoma	Fairly well defined	Hypointense	Heterogeneously intermediate to low signal	Nil (extracompartmental)	Nil	Heterogeneous, small non-enhancing foci

## ○ Carpal tunnel syndrome

- Measure CSA of median nerve at 4 sites
- Proximal border pronator quadratus, proximal to tunnel inlet, at tunnel inlet & at tunnel outlet
- Maximum CSA  $\geq 12 \text{ mm}^2$  at any site  $\rightarrow$  diagnostic of carpal tunnel syndrome
- Maximum CSA = 9-12  $\text{mm}^2 \rightarrow$  borderline
- Maximum CSA  $\leq 9 \text{ mm}^2 \rightarrow$  normal
- Also diagnostic of carpal tunnel syndrome if median nerve CSA in or near carpal tunnel  $> 4 \text{ mm}^2$  median nerve CSA at proximal pronator quadratus level



# 61-year-old man with worsening hip pain

## CASE OUTLINE

Page 1 of 4

## History and CT images

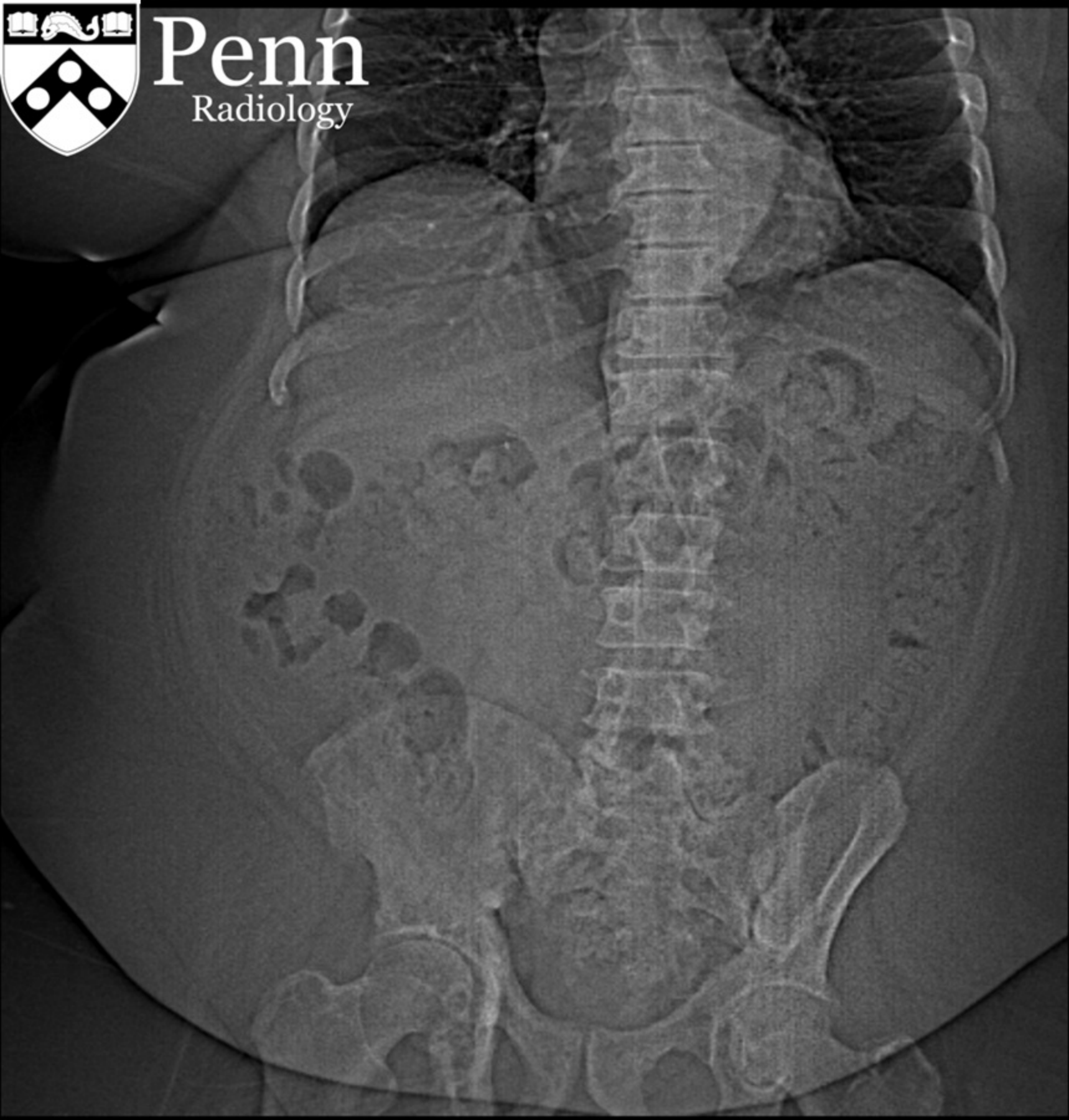
Our appreciation is extended to Dr. Teresa Martin-Carreras, University of Pennsylvania Department of Radiology, for contributing this case.

**History:** A 61-year-old man presented to the emergency department with worsening, chronic, right-sided hip pain, which is worse in the evenings.

An unenhanced CT scan of the abdomen and pelvis was performed. Click images below to enlarge. In order: topogram, axial images in bone window, and coronal and sagittal CT reconstructions in bone window.

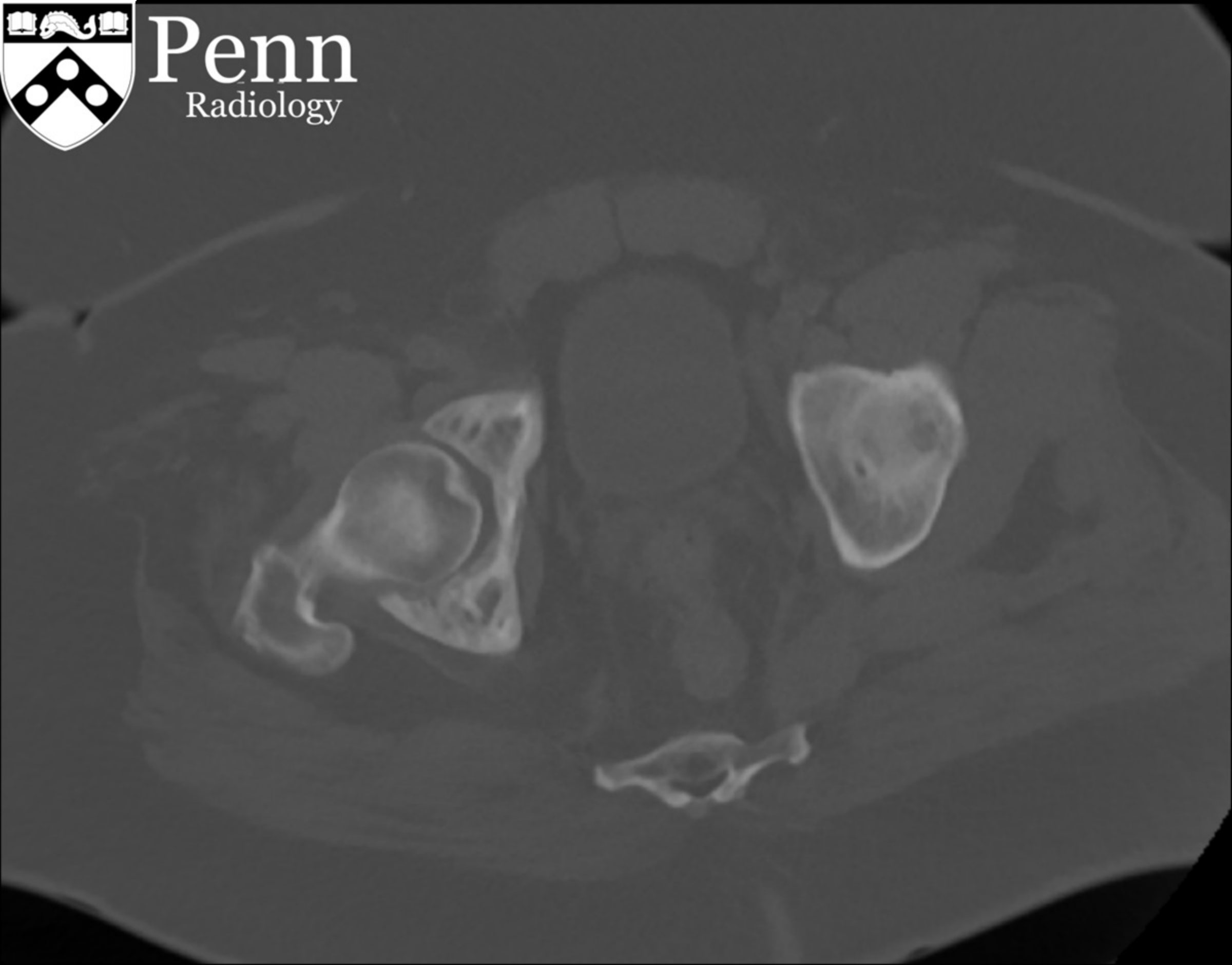


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Radiology



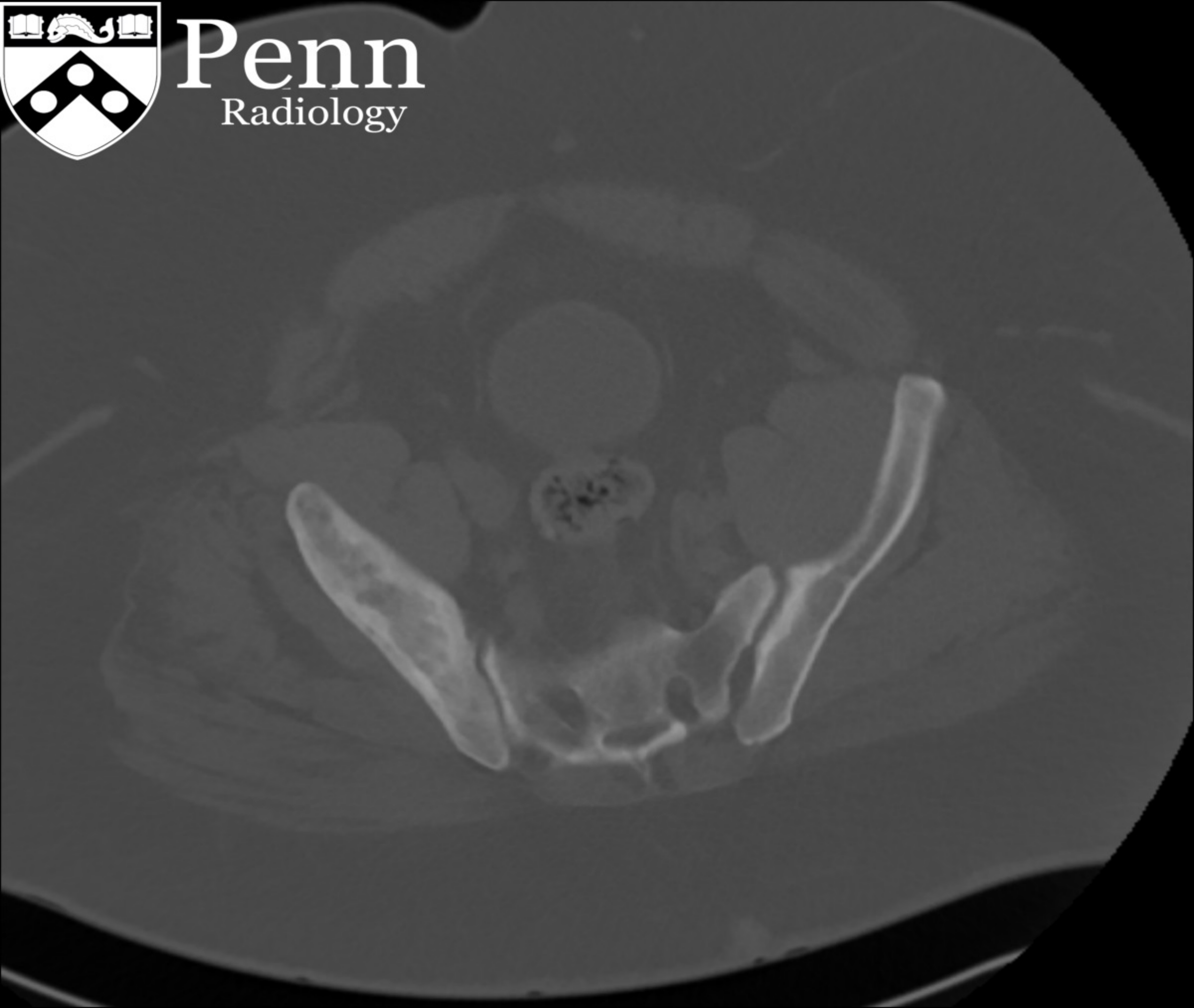


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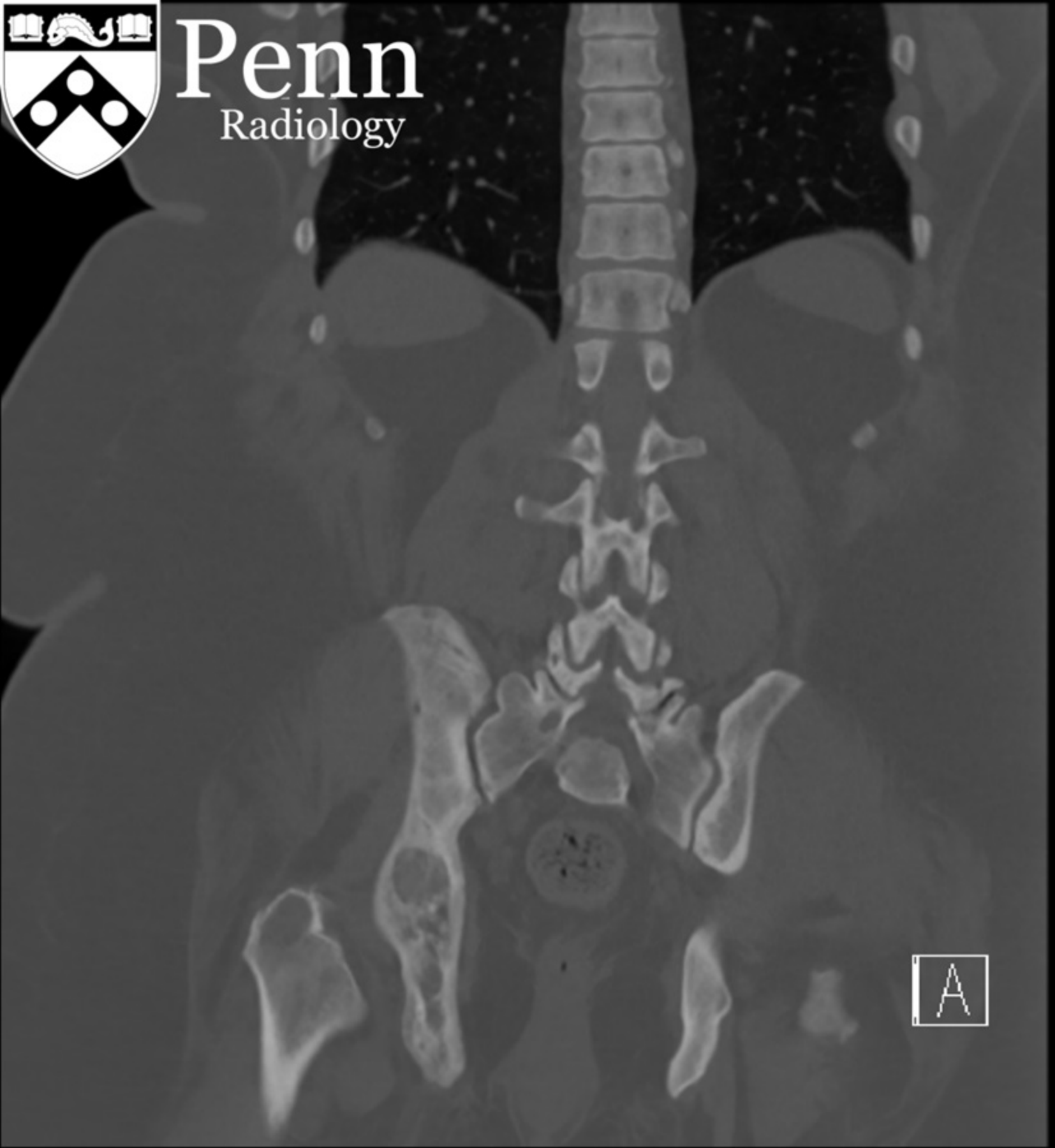


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Radiology





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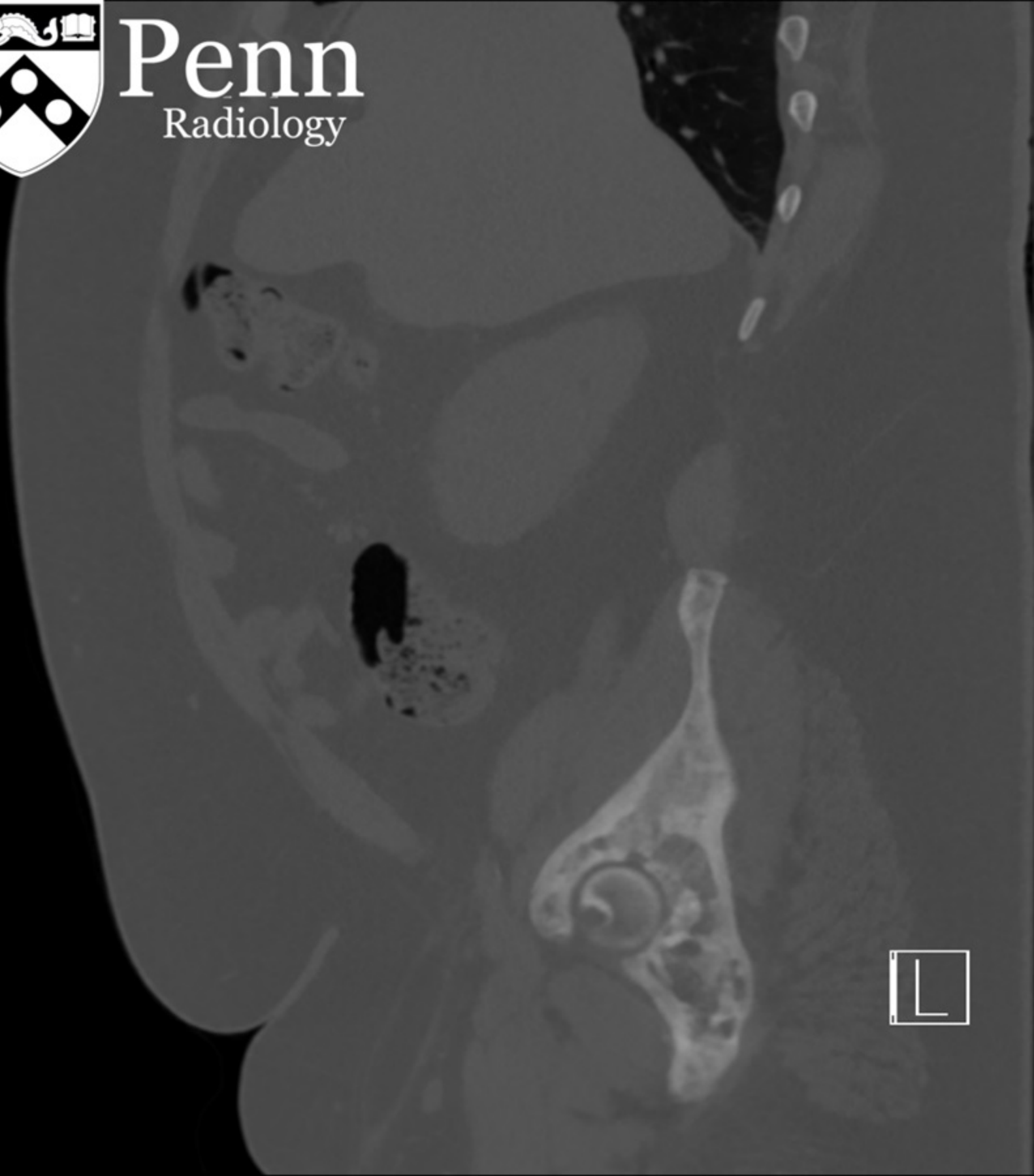


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Radiology



**The salient finding is a normal anatomic variant.**

☐ True

☐ False

The question above accounts for 16% of your total score for this case.

**A fracture is seen on the CT images.**

☐ True

☐ False

**The salient finding is a normal anatomic variant.**

☐ True

☒ False (correct!)

The question above accounts for 16% of your total score for this case.

**A fracture is seen on the CT images.**

☐ True

☒ False (correct!)

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**Sarcomatous degeneration of the process seen in the CT images most commonly results in which of the following?**

- ☐ Chondrosarcoma
- ☐ Malignant fibrous histiocyoma
- ☐ Paget's sarcoma
- ☐ Osteosarcoma

The question above accounts for 16% of your total score for this case.

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- ☐ Chondrosarcoma
- ☐ Malignant fibrous histiocyoma
- ☐ Paget's sarcoma
- ☒ Osteosarcoma (correct!)



The question above accounts for 16% of your total score for this case.

**Which of the following laboratory tests is most likely to be elevated in this patient?**

- ☐ Aspartate transaminase
- ☐ Gamma-glutamyl transpeptidase
- ☐ Alkaline phosphatase
- ☐ Total bilirubin

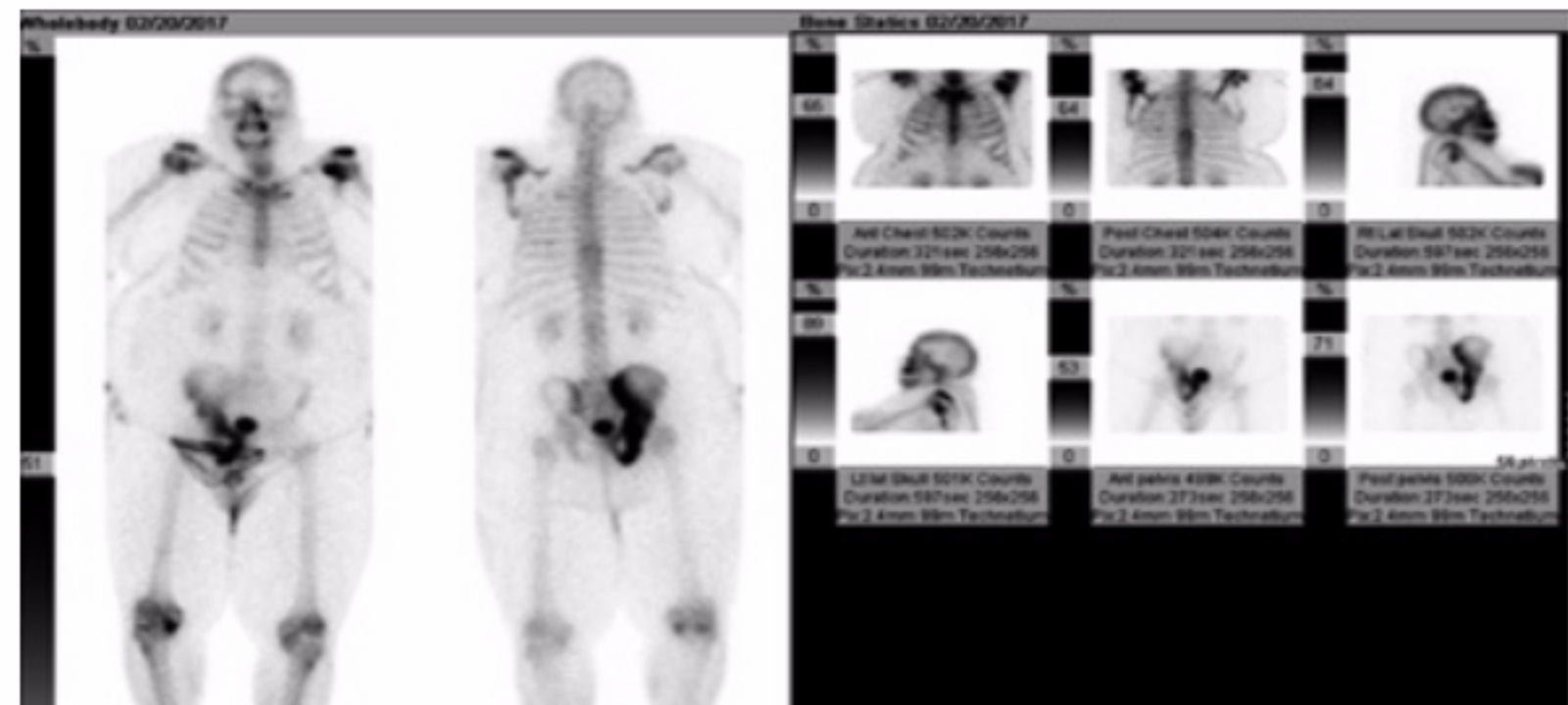
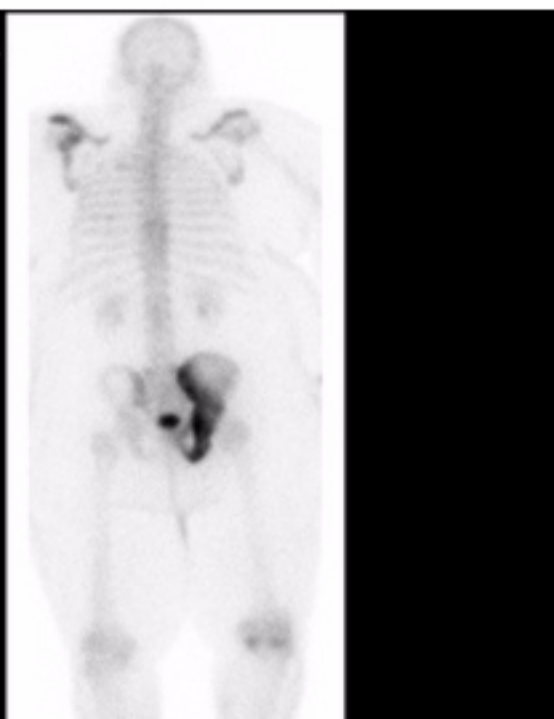
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- ☐ Aspartate transaminase
- ☐ Gamma-glutamyl transpeptidase
- ☒ Alkaline phosphatase (correct!)
- ☐ Total bilirubin

## Additional history and bone scan images

Laboratory workup revealed an isolated elevation of alkaline phosphatase. After review of the CT images and laboratory workup, nuclear medicine bone scintigraphy was performed. Click images below to enlarge. In order: anterior and posterior whole-body images and spot images of the skull, pelvis, and chest.





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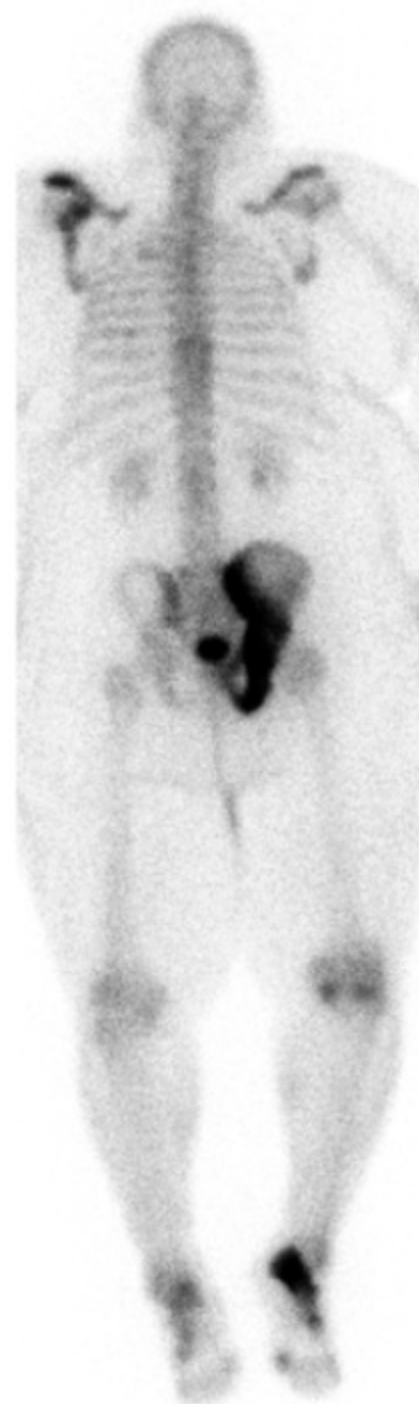




%



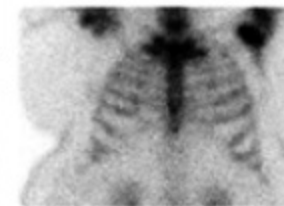
Anterior 99m Technetium



Posterior 99m Technetium

%

65



0

Ant Chest 502K Counts  
Duration:321sec 256x256  
Pix:2.4mm 99m Technetium

%

89

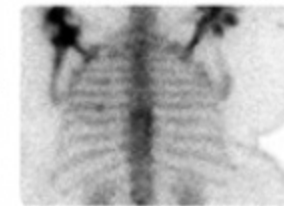


0

Lt lat Skull 501K Counts  
Duration:597sec 256x256  
Pix:2.4mm 99m Technetium

%

64

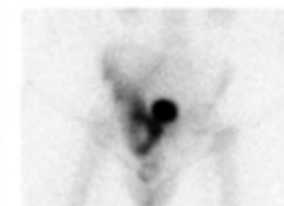


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Post Chest 504K Counts  
Duration:321sec 256x256  
Pix:2.4mm 99m Technetium

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53



0

Ant pelvis 499K Counts  
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%

84

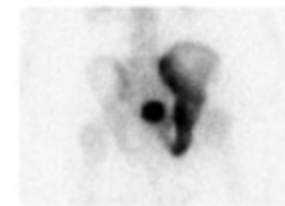


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Rt Lat Skull 502K Counts  
Duration:597sec 256x256  
Pix:2.4mm 99m Technetium

%

71



0

Post pelvis 500K Counts  
Duration:273sec 256x256  
Pix:2.4mm 99m Technetium

50 pixels



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All Images

☐ Technetium-99m diethylenetriamine-pentaacetate (Tc-99m DTPA)

The question above accounts for 16% of your total score for this case.

**What is the most likely diagnosis?**

☐ Fibrous dysplasia

☐ Sclerotic metastases

☐ Multiple myeloma

☐ Bone infarct

☐ Technetium-99m diethylenetriamine-pentaacetate (Tc-99m DTPA)

The question above accounts for 16% of your total score for this case.

**What is the most likely diagnosis?**

☐ Fibrous dysplasia

☐ Sclerotic metastases

☐ Multiple myeloma

☐ Bone infarct

The question above accounts for 10% of your total score for this case.

### What is the most likely diagnosis?

- ☐ Fibrous dysplasia
- ☐ Sclerotic metastases
- ☐ Multiple myeloma
- ☐ Paget's disease of bone

The question above accounts for 20% of your total score for this case.

The question above accounts for 10% of your total score for this case.

### What is the most likely diagnosis?

- ☐ Fibrous dysplasia
- ☐ Sclerotic metastases
- ☐ Multiple myeloma
- ☒ Paget's disease of bone (correct!)

The question above accounts for 20% of your total score for this case.



## Findings

- **CT:** CT demonstrates an asymmetric enlargement and sclerosis of the right iliac wing with coarsened trabeculae. No evidence of fracture is seen. There are multilevel degenerative changes in the visualized portions of the thoracolumbar spine.
- **Bone scan:** There is increased tracer uptake in the right hemipelvis. Increased tracer uptake around both shoulders (left more than right), knees, ankles, and right foot are favored to be degenerative changes. Tracer distribution is otherwise physiologic. Both kidneys are visualized.

## Differential diagnosis

- Paget's disease
- Sclerotic metastases
- Fibrous dysplasia

## Differential diagnosis

- Paget's disease
- Sclerotic metastases
- Fibrous dysplasia
- Multiple myeloma
- Myelofibrosis

**Diagnosis:** Paget's disease of the right hemipelvis

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# Paget's disease of bone

## Pathophysiology

- Paget's disease of bone is also known as osteitis deformans.
- It refers to a disease of disordered and increased bone turnover and remodeling.
- Patients have enlarged bone with coarsened trabeculae in the involved area.
- The condition can be monostotic (10% to 35% of cases) or polyostotic (65% to 90% of cases).
- The distribution of Paget's disease of bone is variable.
  - Skull involvement: 25% to 65%
  - Proximal long bones: 25% to 30%
  - Pelvis: 30% to 75%
  - Spine: 30% to 75%
- Sarcomatous degeneration may be due to high rates of bone turnover leading to mutations and genomic deletions. Incidence of sarcomatous change is 0.7% to 0.95%.



- Sarcomatous degeneration may be due to high rates of bone turnover leading to mutations and genomic deletions. Incidence of sarcomatous change is 0.7% to 0.95%.
- The most common distribution of Paget's sarcoma is listed below:
  - Two-thirds occur in large limb bones (i.e., femur, tibia, humerus).
  - One-third occurs in flat bones (i.e., pelvis, scapula, skull).
  - Multifocal Paget's sarcoma is usually seen in the femur and skull.
- Most commonly, however, Paget's disease degenerates to osteosarcoma (50% to 60%), chondrosarcoma, or malignant fibrous histiocytoma.
- A genetic component linked to chromosome arm 18q may predispose patients to Paget's disease.
- Mutations in gene encoding sequestosome 1 (SQSTM1) also have been found in familial (25% to 50%) and sporadic Paget's disease.
- Lytic, intermediate, and sclerotic stages have been described.
  - Lytic stage: Active, osteoclasts line trabeculae, with increased vascularity.
  - Intermediate stage: Active, osteoclasts and osteoblasts line seams of osteoid.
  - Sclerotic stage: Late inactive, osteoblasts predominate, reactive sclerosis with

- Lytic, intermediate, and sclerotic stages have been described.
  - Lytic stage: Active, osteoclasts line trabeculae, with increased vascularity.
  - Intermediate stage: Active, osteoclasts and osteoblasts line seams of osteoid.
  - Sclerotic stage: Late inactive, osteoblasts predominate, reactive sclerosis with thickened trabeculae, and no remodeling of incomplete insufficiency fractures.
- Elevated alkaline phosphatase and elevated urinary hydroxyproline may be noted on laboratory workup.

## Epidemiology

- There is a slight male predilection.
- Generally seen in patients 55 to 85 years old.
- There is a Caucasian predominance, especially in the U.K., Australia, U.S., New Zealand, and Western Europe.
- Paget's sarcoma is more common in men (2:1), with median age of 64 years.
- An environmental (viral) etiology has been proposed due to the regional difference in



disease prevalence.

- May be associated with chronic measles.
- Intranuclear inclusion bodies seen in osteoclasts resembling those of *Paramyxovirus* have been identified.

## Clinical presentation

- Patients may be asymptomatic for many years.
- They may present with deep, constant bone pain that is worse at night.
- They may have deformities (i.e., protrusio, femoral or tibial bowing).
- Patients can present with increasing skull size, hearing loss (cranial nerve VIII impingement), and/or spinal stenosis.
- In 12% to 20% of cases, patients will see pathologic fractures, most commonly involving the femur.
- Patients can present with Paget's sarcoma, clinically described as a change in pain pattern.

- Patients can present with Paget's sarcoma, clinically described as a change in pain pattern.
- Patients may complain of warm skin due to increased blood flow to the affected bone.
- Rarely, patients may present with high-output cardiac failure.

## Imaging features

- Radiographs:
  - Early lesions will show a lytic, thinned cortex, while later lesions will have mixed lytic/sclerotic components.
  - Disordered, thickened trabeculae with enlargement of the involved bone in all dimensions will be seen.
  - Numerous deformities related to bone softening (e.g., protrusio, varus hips, anterior bowing tibia, basilar invagination) may be seen.
  - In the skull, enlargement or lytic lesions termed osteoporosis circumscripta may be seen. In later phases, may see focal regions of sclerosis, termed cotton wool.



- The long bones may show a sharp oblique delineation at the border with normal bone termed blade of grass or have a flame-shaped appearance.
- An incomplete horizontal insufficiency fracture may be seen on the convex side of the bone which may progress to “banana” fractures.
- In the pelvis, early cortical thickening and sclerosis of iliopectineal and ischiopubic lines are seen, as well as enlargement/involvement of the iliac wing. These findings are often asymmetrical and more commonly occur on the right.
- In the spine, enlarged “picture frame” sign is seen as enlarged vertebrae with thickened sclerotic borders and radiolucent centers. “Ivory vertebra” sign in late blastic phase also may be seen.
- Sarcomatous degeneration will be seen as a permeative process with cortical breakthrough and a soft-tissue mass component.
- CT:
  - CT is very useful for evaluating the skull and spine.
  - Temporal bone thickening and also enlargement and sclerosis of the base of the skull with diploic space widening will be seen.

- Temporal bone thickening and also enlargement and sclerosis of the base of the skull with diploic space widening will be seen.
- CT allows for better assessment of spinal stenosis.
- MRI:
  - Appearance with MRI will be variable, depending on the phase of the disease.
  - Often contains more marrow fat than adjacent normal bone.
  - Often heterogeneous "speckled" on both T1- and T2-weighted images.
  - Inhomogeneous enhancement is often seen.
  - Sarcomatous degeneration will be seen as an aggressive soft-tissue mass, and there will often be marrow fat obliteration by a superimposed tumor. The absence of fat may help differentiate tumor from Paget's disease of bone.
  - Dynamic contrast imaging may show regions of excessive hypervascularity and may help evaluate the effect of medications.
- Tc-99m MDP bone scintigraphy:
  - Tc-99m MDP bone scintigraphy has a high sensitivity but is not specific.
  - It will show marked increased uptake in all phases of the disease, although uptake



- Tc-99m MDP bone scintigraphy has a high sensitivity but is not specific.
- It will show marked increased uptake in all phases of the disease, although uptake may be normal in the burnt-out sclerotic quiescent phase.

## Treatment

- No treatment is warranted if the patient is asymptomatic.
- Medical treatment options include bisphosphonates, calcitonin, and mithramycin.
- In some instances, surgery (arthroplasty) may be performed to correct deformities.
- In the case of Paget's sarcoma, treatment depends on whether the disease is metastatic at presentation.
  - Aggressive treatment includes systemic chemotherapy, radiation, and wide resection.
  - Palliative treatment includes stabilization of involved bone and systemic chemotherapy as necessary.



## 56-year-old man with neuroendocrine tumor, intractable pain

### CASE OUTLINE

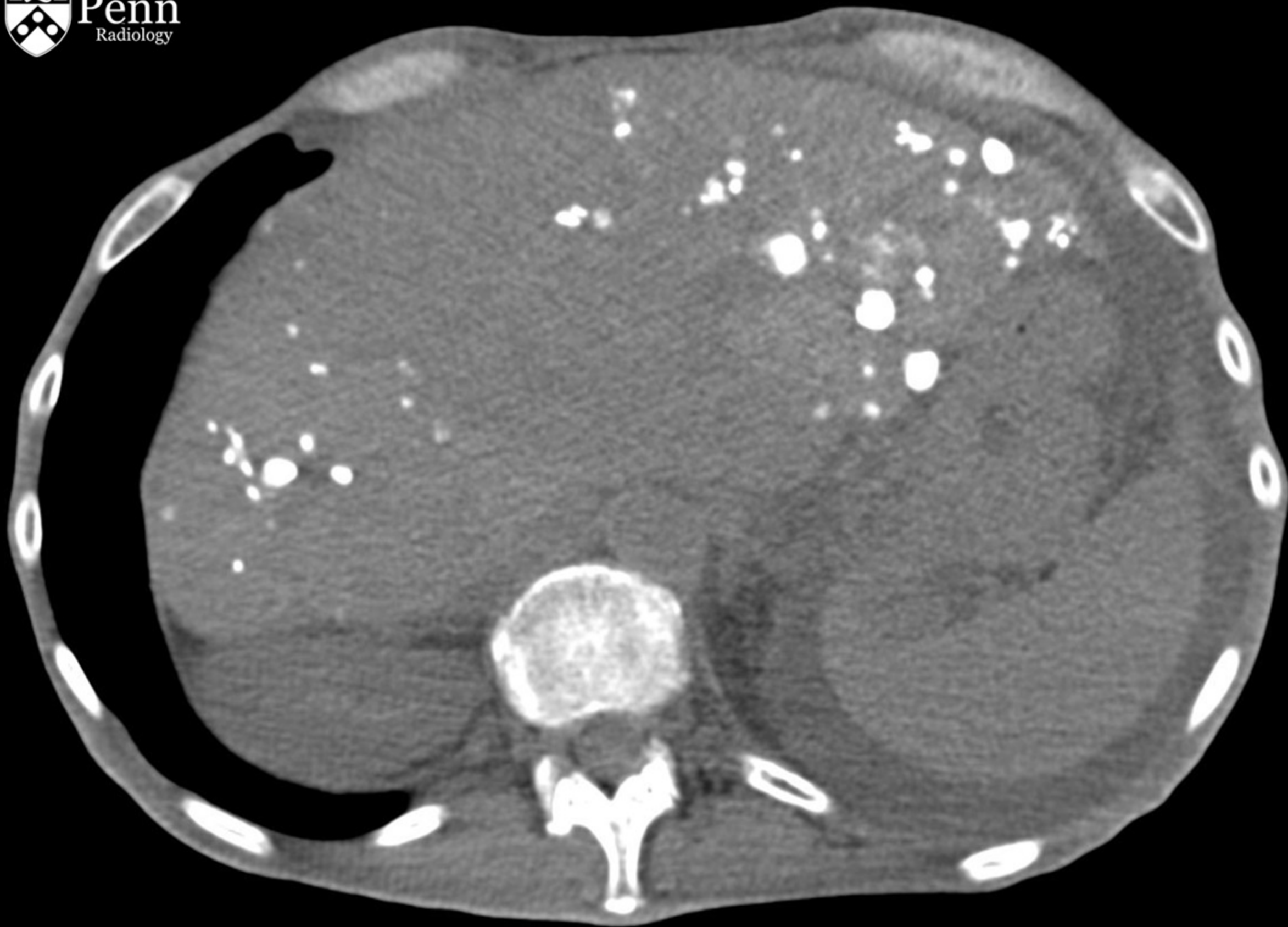
Page 1 of 4

## History and CT images

Our appreciation is extended to Dr. James X. Chen, University of Pennsylvania Department of Radiology, for contributing this case.

**History:** A 56-year-old man with a metastatic low-grade pancreatic neuroendocrine tumor presents with several months of intractable abdominal pain.

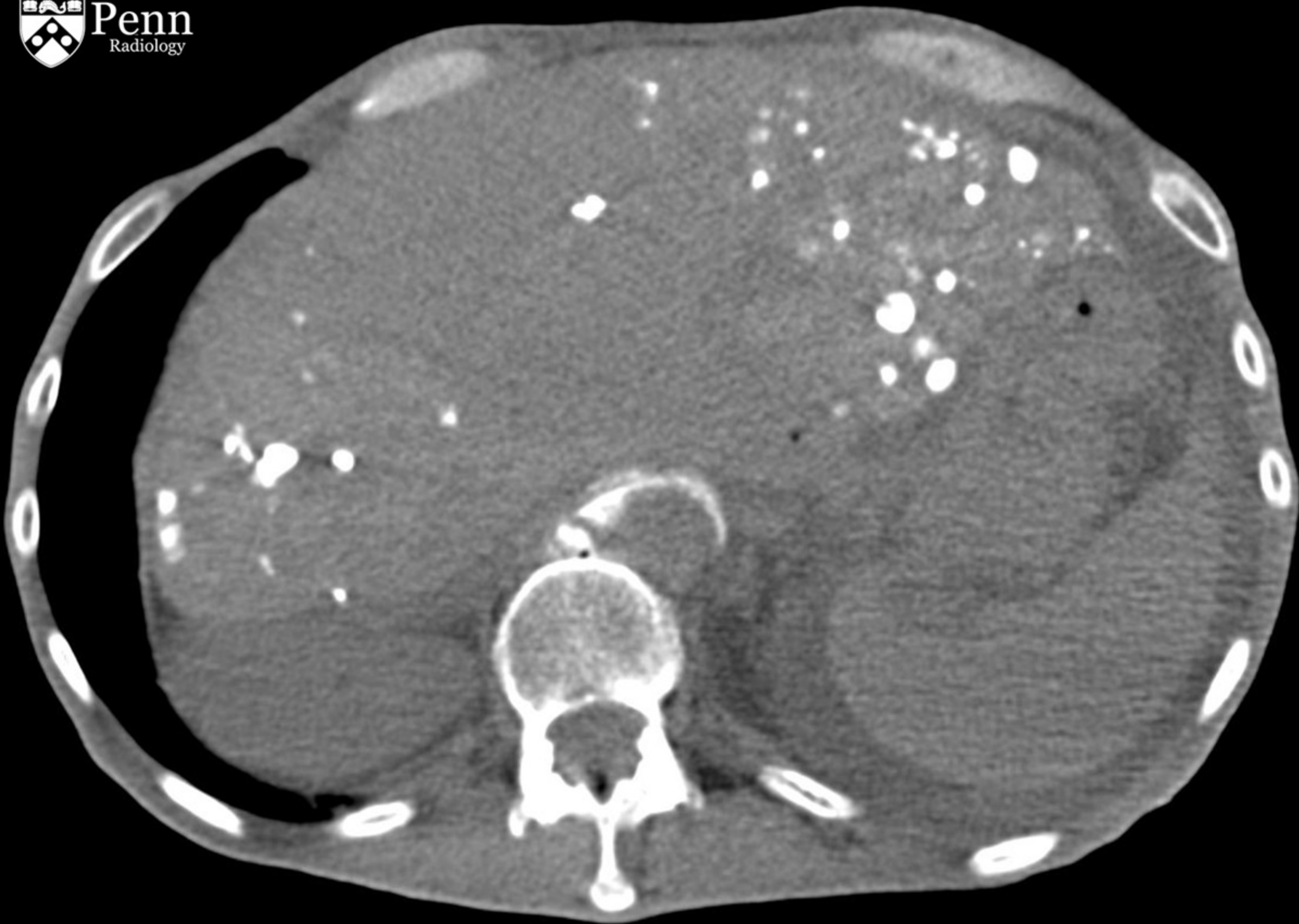
A CT-guided procedure was performed. Click images below to enlarge.













## What is the most likely etiology of the hyperattenuating lesions within the liver?

- ☐ Chronic portal vein thrombosis
- ☐ Lipiodol from prior chemoembolization
- ☐ Polycystic liver disease
- ☐ Echinococcal cysts
- ☐ Hepatic tuberculosis

**What is the most likely etiology of the hyperattenuating lesions within the liver?**

- ☐ Chronic portal vein thrombosis
- ☒ Lipiodol from prior chemoembolization (correct!)
- ☐ Polycystic liver disease
- ☐ Echinococcal cysts
- ☐ Hepatic tuberculosis

**Which of the following anatomic approaches has been used for the above procedure?**

☐ Anterior

☐ Posterior

☐ Lateral decubitus

☐ None of the above

The question above accounts for 13% of your total score for this case.

**Which of the following anatomic approaches has been used for the above procedure?**

☒ Anterior (correct!)

☐ Posterior

☐ Lateral decubitus

☐ None of the above

The question above accounts for 13% of your total score for this case.

**What procedure is demonstrated in the above images?**

☐ Cryoablation of adrenal metastases

☐ Portal vein embolization

☐ Celiac plexus neurolysis

☐ Retroperitoneal lymph node biopsy

The question above accounts for 13% of your total score for this case.



**What procedure is demonstrated in the above images?**

- ☐ Cryoablation of adrenal metastases
- ☐ Portal vein embolization
- ☒ Celiac plexus neurolysis (the correct answer)
- ☐ Retroperitoneal lymph node biopsy (wrong) <- your answer

The question above accounts for 13% of your total score for this case.

**Which of the following is NOT administered as part of the above procedure?**

☐ Lipiodol

☐ Iodinated contrast

☐ Bupivacaine

☐ Phenol

The question above accounts for 13% of your total score for this case.

**Which of the following is NOT administered as part of the above procedure?**

☒ Lipiodol (correct!)

☐ Iodinated contrast

☐ Bupivacaine

☐ Phenol

The question above accounts for 13% of your total score for this case.

Prior images demonstrated CT-guided celiac plexus neurolysis from an anterior transhepatic approach.

**Which of the following is the usual level of the celiac plexus?**

☐ T8-T9

☐ T10-T11

☐ T12-L1

☐ L2-L3

Prior images demonstrated CT-guided celiac plexus neurolysis from an anterior transhepatic approach.

**Which of the following is the usual level of the celiac plexus?**

☐ T8-T9

☐ T10-T11

☒ T12-L1 (correct!)

☐ L2-L3



**Which of the following has been associated with improved efficacy of pain control from celiac plexus neurolysis?**

- ☐ Administration of a lower volume of neurolytic
- ☐ Greater extent of diffusion of the neurolytic agent along the antecrural space
- ☐ Use of an anterior approach as compared to a posterior approach
- ☐ Patient dependency on higher opioid doses prior to the procedure

The question above accounts for 12% of your [total score for this case](#)

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The question above accounts for 12% of your [total score for this case](#)

The question above accounts for 12 % of your total score for this case.

**Which of the following is an advantage of using phenol over ethanol as the neurolytic agent?**

- ☐ Superior efficacy for pain control
- ☐ Decreased procedural pain
- ☐ Lower cost
- ☐ Improved diffusion in the antecrural space

The question above accounts for 12 % of your total score for this case.

**Which of the following is an advantage of using phenol over ethanol as the neurolytic agent?**

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- ☐ Lower cost
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The question above accounts for 12% of your total score for this case.



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**Which of the following is a side effect of celiac axis neurolysis?**

- ☐ Back pain
- ☐ Orthostatic hypotension
- ☐ Transient diarrhea
- ☐ All of the above

The question above accounts for 12% of your total score for this case.



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- ☐ Back pain
- ☐ Orthostatic hypotension
- ☐ Transient diarrhea
- ☒ All of the above (correct!)

The question above accounts for 12% of your total score for this case.

## Findings

Preliminary CT image obtained through the upper abdomen demonstrates hyperattenuating foci through the liver (image 1), corresponding to lipiodol staining from prior transarterial chemoembolization for liver metastases from the pancreatic neuroendocrine tumor. Using an anterior approach, two 22-gauge Chiba needles were advanced through the liver in the antecrural and right retrocrural spaces at the level of the celiac axis (image 2). Iodinated contrast was injected through the needles to confirm satisfactory tip position (image 3), followed by a test injection of bupivacaine to assess for adequate analgesia. Then, a mixture of iodinated contrast and phenol was infused, and a postinfusion scan was performed to confirm free diffusion of the solution along the antecrural and retrocrural spaces (image 4).

**Differential diagnosis** (other image-guided interventions performed in this anatomic region)

performed to confirm free diffusion of the solution along the antecrural and retrocrural spaces (image 4).

**Differential diagnosis** (other image-guided interventions performed in this anatomic region)

- Celiac plexus neurolysis
- Cryoablation adrenal metastases
- Retroperitoneal lymph node biopsy

**Diagnosis:** CT-guided celiac plexus neurolysis for intractable malignancy-associated abdominal pain

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# Celiac plexus neurolysis

## Pathophysiology

- Abdominal pain can be debilitating in patients with abdominal malignancies. Pain management often requires high-dose narcotics, which have side effects that reduce quality of life.
- Celiac plexus:
  - Largest visceral plexus, located in the retroperitoneum along the anterolateral aorta, at the level of the celiac trunk and superior mesenteric artery (typically T12-L1).
  - Consists of an interconnected network of nerve fibers, including the following:
    - Visceral afferent fibers: Responsible for nociception from upper abdominal visceral organs.
    - Preganglionic sympathetic and parasympathetic efferent nerve fibers
  - Disruption of these nerves can provide pain relief from intractable malignancy-

- Disruption of these nerves can provide pain relief from intractable malignancy-associated abdominal pain.

## **Imaging features**

Unenhanced CT is the preferred modality for guidance in percutaneous celiac plexus block/neurolysis.

- Paired structure:
  - Left ganglion is visible in approximately 90% cases -- located between the adrenal gland and diaphragmatic crus.
  - Right ganglion is visible in approximately 67% cases -- located between right diaphragmatic crus and inferior vena cava.
- Attenuation is similar to adrenal glands.
- Morphology is multilobulated or discoid.

## **Treatment**



## Treatment

- Celiac plexus neurolysis is the permanent disruption of the celiac plexus with phenol (less painful) or ethanol.
- Indications:
  - Intractable abdominal pain from upper abdominal primary malignancies (biliary, esophageal, gastric, pancreatic) or liver metastases.
  - Also improves nausea in patients with pancreatic cancer by increasing gastric motility (due to sympathetic denervation).
- Contraindications:
  - Uncorrectable coagulopathy
  - Abdominal aortic aneurysm
  - Eccentric origin of the celiac artery
  - Local infection or sepsis
- Agents:
  - Ethanol causes transient, severe pain and is typically injected as a mixture with

- Agents:
  - Ethanol causes transient, severe pain and is typically injected as a mixture with bupivacaine for analgesia.
  - Phenol is less painful than ethanol but may have less efficacy.
- Injection sites:
  - Antecrural: Anterior to the crus and aorta and directly destroys the celiac plexus.
  - Retrocrural: Posterior to the crus and destroys the splanchnic nerves.
- Approaches:
  - Prone, supine, lateral decubitus, and oblique positions can be used.
    - Prone position is preferred.
    - Anterior approach often requires traversal of visceral organs, which usually does not result in any sequelae due to the small gauge of the access needle. Traversal of the pancreas is associated with the risk of pancreatitis.
  - A thin (20- to 24-gauge) Chiba needle is advanced 1-2 cm anteriorly to the aorta.
  - After aspirating to confirm no blood return, dilute iodinated contrast is injected and should diffuse freely in the antecrural space.



should diffuse freely in the antecrural space.

- Local anesthetic (bupivacaine) is then injected to test the efficacy of the analgesia prior to permanent neurolysis.
- Neurolytic agent is injected last.
- A CT scan is performed to demonstrate diffusion of the injected agent along the retroperitoneum and anterolateral aortic wall.
- Complications:
  - Back pain -- most common
  - Orthostatic hypotension (up to 50% of patients) – results from decreased sympathetic tone
  - Transient diarrhea (between 40% to 50% of patients) – increased peristalsis from unopposed parasympathetic activity
- Efficacy:
  - Pain control is generally more effective when treatment is performed in earlier-stage disease and in patients with lower opioid requirements.
  - Most important technical determinants for effective pain control are the amount of

unopposed parasympathetic activity

- Efficacy:
  - Pain control is generally more effective when treatment is performed in earlier-stage disease and in patients with lower opioid requirements.
  - Most important technical determinants for effective pain control are the amount of neurolytic administered and the degree of diffusion of the agent along the antecrural space

## References

1. Kambadakone A, Thabet A, Gervais DA, Mueller PR, Arellano RS. CT-guided celiac plexus neurolysis: A review of anatomy, indications, technique, and tips for successful treatment. *Radiographics*. 2011;31(6):1599-1621.
2. Koyyalagunta D, Engle MP, Yu J, Feng L, Novy DM. The effectiveness of alcohol versus phenol based splanchnic nerve neurolysis for the treatment of Intra-abdominal cancer pain. *Pain Physician*. 2016;18(4):281-292.



## 61-year-old man with posterior heel pain

### CASE OUTLINE

Page 1 of 4

## History and radiograph

Our appreciation is extended to Dr. Akash Patel, University of Pennsylvania Department of Radiology, for contributing this case.

**History:** A 61-year-old man presents with progressive, sharp, shooting posterior heel pain of the right foot for the past two months.

Lateral radiograph of the right foot is shown below. Click to enlarge.



R  
KS

5 cm



**There is a bony abnormality involving which of the following calcaneal landmarks?**

- ☐ Calcaneal tubercle
- ☐ Posterosuperior osseous protuberance of the calcaneus
- ☐ Calcaneal sulcus
- ☐ Talar articular surface

The question above accounts for 16% of your total score for this case.

**There is a bony abnormality involving which of the following calcaneal landmarks?**

☐ Calcaneal tubercle

☒ Posterosuperior osseous protuberance of the calcaneus (correct!)

☐ Calcaneal sulcus

☐ Talar articular surface

The question above accounts for 16% of your total score for this case.

The question above accounts for 16% of your total score for this case.

**What is the name of this osseous deformity?**

☐ Charcot's deformity

☐ Jones' deformity

☐ Haglund's deformity

The question above accounts for 16% of your total score for this case.

**This deformity may be seen in asymptomatic patients**

The question above accounts for 16% of your total score for this case.

**What is the name of this osseous deformity?**

☐ Charcot's deformity

☐ Jones' deformity

☒ Haglund's deformity (correct!)

**[Explain this Answer]**

The question above accounts for 16% of your total score for this case.



**This deformity may be seen in asymptomatic patients.**

☐ True

☐ False

The question above accounts for 17% of your total score for this case.

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1

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[next »](#)

**This deformity may be seen in asymptomatic patients.**

☒ True (correct!)

☐ False

**[Explain this Answer]**

The question above accounts for 17% of your total score for this case.

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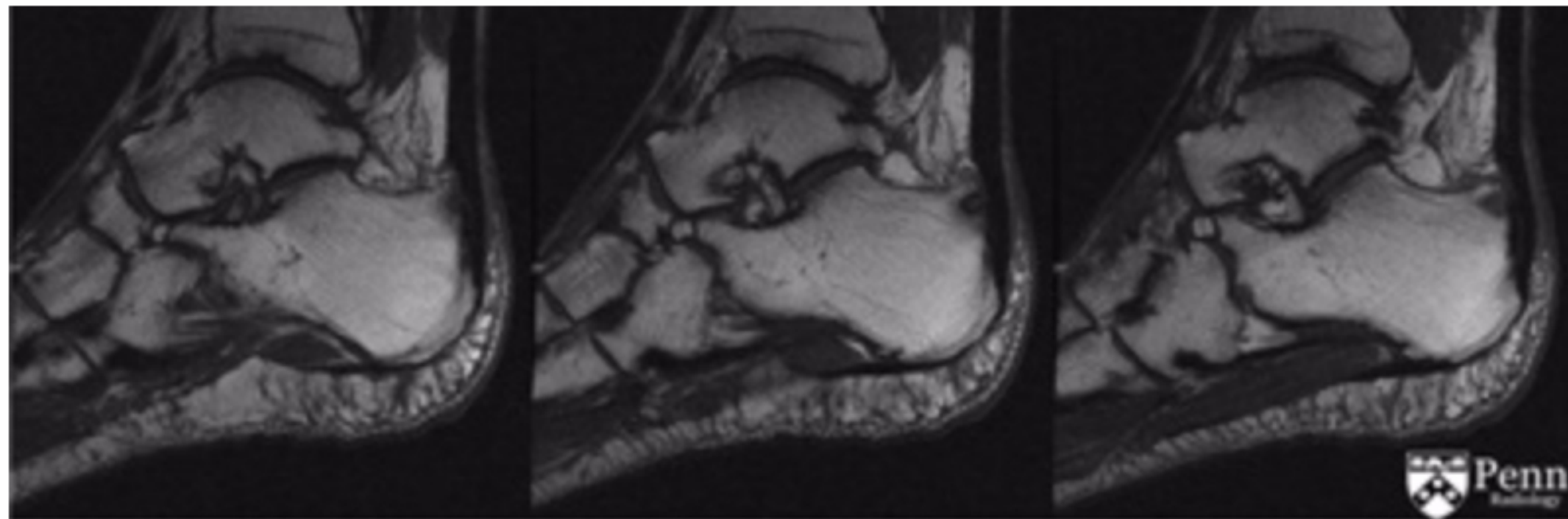
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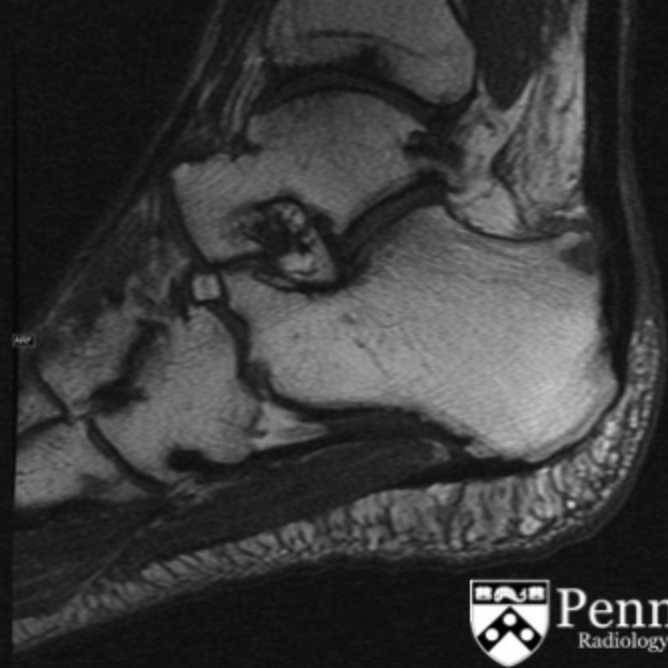
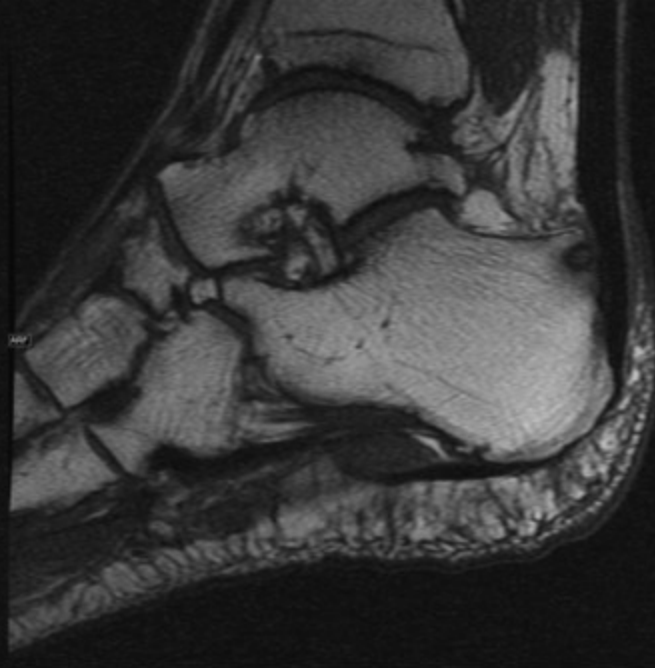
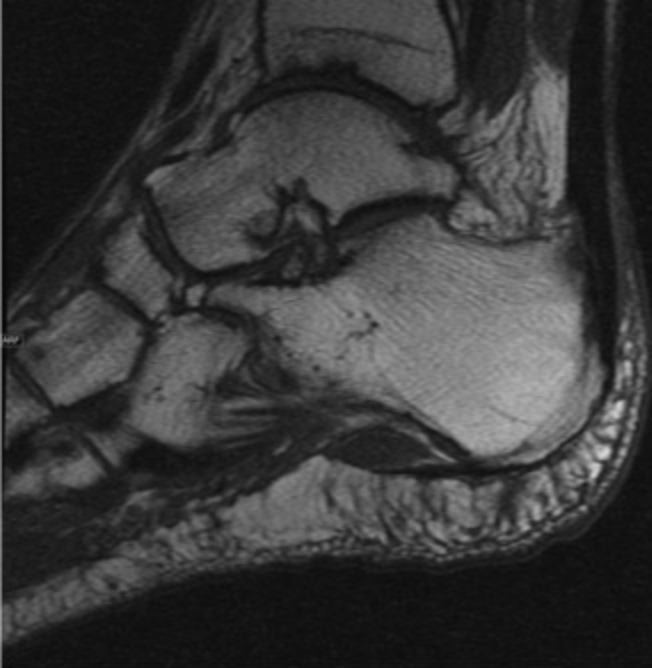
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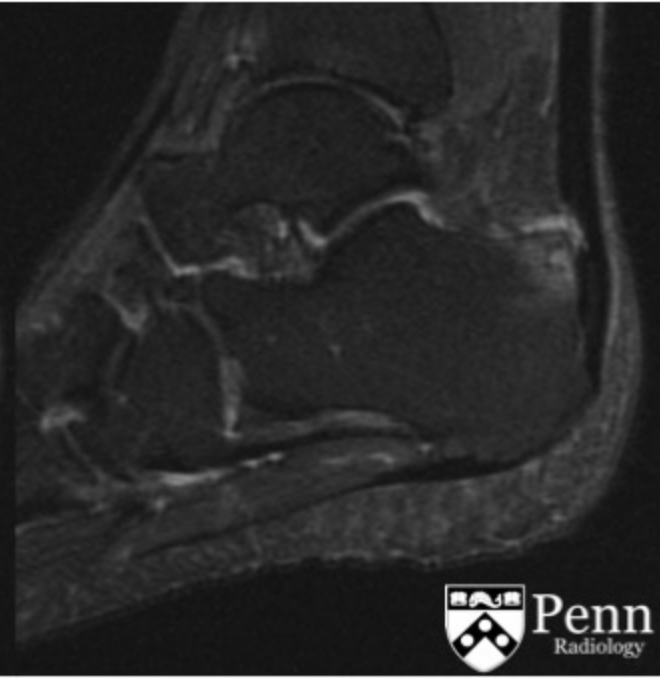
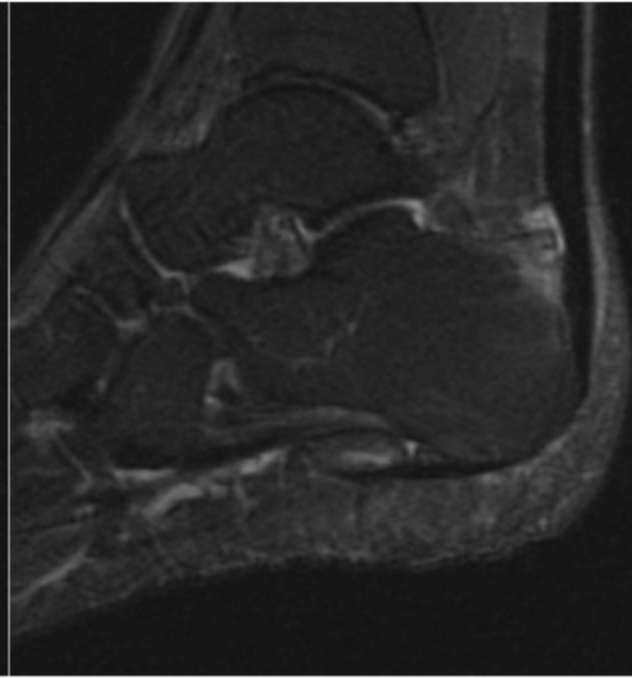
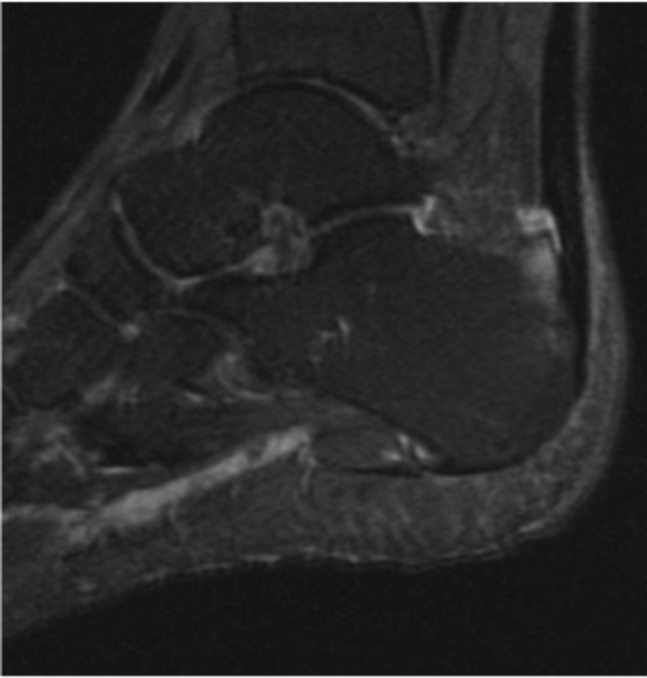
[next »](#)

## MR images

Sagittal T1-weighted and proton density-weighted fat-saturated MR images are shown below. Click to enlarge.









**The Achilles tendon is intact.**

☐ True

☐ False

The question above accounts for 17% of your total score for this case.

**Which of the following findings is noted on MRI?**

☐ Achilles tendon insertional tendinopathy

☐ Partial thickness tear

**The Achilles tendon is intact.**

☐ True

☒ False (correct!)

**[Explain this Answer]**

The question above accounts for 17% of your total score for this case.

**Which of the following findings is noted on MRI?**

☐ Achilles tendon insertional tendinopathy



False. There is a partial tear of the Achilles near its origin.

## The Achilles te

☐ True

☒ False (correct)

[[Explain this Answer](#)]

The question above

Which of the fo

## Which of the following findings is noted on MRI?

- ☐ Achilles tendon insertional tendinopathy
- ☐ Retrocalcaneal bursitis
- ☐ Trace fluid posterior to the Achilles tendon
- ☐ Bony edema of the posterosuperior prominence of the calcaneus
- ☐ All of the above

## Which of the following findings is noted on MRI?

- ☐ Achilles tendon insertional tendinopathy
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- ☐ Trace fluid posterior to the Achilles tendon
- ☐ Bony edema of the posterosuperior prominence of the calcaneus
- ☒ All of the above (correct!)



**Which of the following is a potential treatment option for this patient?**

☐ A. Reassessment of footwear and use of heel inserts

☐ B. Oral anti-inflammatory medications

☐ C. Surgical retrocalcaneal decompression and calcaneal ostectomy/osteotomy

☐ D. All of the above

☐ E. Both A and B

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☒ D. All of the above (correct!)

☐ E. Both A and B

## Findings

- **Radiograph:**

- Prominence of the posterosuperior osseous protuberance of the calcaneus (at the insertion of the Achilles tendon) is seen.
- There is nonvisualization of the normal retrocalcaneal recess -- suggestive of retrocalcaneal bursitis.
- There is subtle localized swelling and convexity of the soft tissues posterior to the insertion site of the Achilles tendon.

- **MRI:**

- Haglund's deformity is seen with associated bony edema.
- Mild insertional tendinopathy of the Achilles tendon with a partial tear near its insertion site.
- Minimal fluid behind the Achilles tendon, which may represent mild retro-Achilles bursitis.

bursitis.

## Differential diagnosis

- Haglund's syndrome
- Rheumatoid arthritis
- Reiter's syndrome
- Stress fracture of the posterior calcaneal process

**Diagnosis:** Haglund's syndrome

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# Haglund's deformity/syndrome

## Pathophysiology

- Haglund's deformity is the prominent posterosuperior osseous protuberance of the calcaneus.
- Haglund's syndrome is a constellation of findings and bony abnormalities representing one cause of retrocalcaneal pain due to inflammation of regional soft tissues.
  - Retrocalcaneal bursitis
  - Superficial tendo-Achilles bursitis
  - Thickening or inflammation of the Achilles tendon
  - Prominent bony contour of the posterior calcaneus
- A rigid and plantarflexed first toe may predispose a patient for a Haglund's deformity and retrocalcaneal bursitis due to varus deformity of the hindfoot.
- Prominent plantar osseous projections also can alter the bone-soft tissue interface of the hindfoot.



- Additional biomechanical risk factors for Haglund's syndrome include the following:
  - Rear foot equinus
  - Compensated rearfoot varus
  - Compensated forefoot valgus
  - Cavus deformity
  - Trauma to apophysis in childhood

## **Epidemiology**

Haglund's deformity is associated with young women who wear "pump-style" shoes, hence the nickname "pump bump."

## **Clinical presentation**

- Pain over the posterior calcaneal tuberosity
- Palpable soft-tissue prominence in this region (pump bump)
- Pain with flexion of foot

- Palpable soft-tissue prominence in this region (pump bump)
- Pain with flexion of foot
- Prominent calcaneal posterosuperior protuberance, usually on lateral side, often with local swelling and inflammatory changes
- Haglund deformity: often found in asymptomatic patients
- Haglund's syndrome: may be present even without the deformity

## Imaging features

- All modalities: Haglund deformity appears as a prominent posterosuperior osseous protuberance of the calcaneus (at the insertion of the Achilles tendon).
  - Radiographic angles have been described but do not correlate well with symptoms.
  - There are no truly validated criteria for the enlarged prominence.
- Radiographs/CT:
  - Retrocalcaneal bursitis -- nonvisualization of the normal retrocalcaneal recess

- Radiographs/CT:
  - Retrocalcaneal bursitis -- nonvisualization of the normal retrocalcaneal recess
    - The normal retrocalcaneal recess extends 2 mm below the bursal projection and has a sharp interface along the anterior edge of the Achilles tendon.
  - Achilles tendinitis -- thickening (anteroposterior diameter more than 9 mm, 2 cm above the bursal projection) and loss of the sharp interface
  - Superficial tendo-Achilles bursitis -- localized swelling and convexity of the soft tissues posterior to the insertion site of the Achilles tendon
    - Superficial tendo-Achilles bursa forms in response to chronic trauma and is posterior to the Achilles tendon insertion.
- MRI:
  - Retrocalcaneal bursitis
  - Achilles tendinitis
  - Superficial tendo-Achilles bursitis
- Ultrasound:
  - Fluid in pre- or retro-Achilles bursae



- Ultrasound:
  - Fluid in pre- or retro-Achilles bursae
    - Note: A very small amount of fluid may be seen in normal patients.
  - Thickening and heterogeneity of the insertion of the Achilles tendon

## Differential diagnoses

- Rheumatoid arthritis
- Reiter's syndrome
- Achilles insertional tendinopathy
- Stress fracture of the posterior calcaneal process
- Accessory soleus

## Treatment

- Conservative treatment options include the following:

Reassessment of footwear

## Treatment

- Conservative treatment options include the following:
  - Reassessment of footwear
  - Use of heel inserts
  - Oral anti-inflammatory medication
- Steroid injection into pre-Achilles bursa
- Surgical treatment: Options include retrocalcaneal decompression and calcaneal ostectomy/osteotomy.
  - Patients may have a recurrence of symptoms with inadequate bone resection.
  - Weakening of the gastrocnolius complex may occur with excessive bone resection.

## References

1. Crim JR. Haglund syndrome. STATdx.com. Accessed February 22, 2017.